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Fibrous Dysplasia-A Review

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Abstract: Fibrous dysplasia lesions is a descriptive term used to denote lesions that demonstrate replacements of normal bone by a fibrous connective tissue matrix, within which varying amounts of immature and mature bone & in some instances, cementous like tissue are deposited. It is described as one of the most perplexing diseases of osseous tissue of unknown etiology, uncertain pathology, and diverse histopathology. Fibrous dysplasia is a developmental tumor-like condition or as considered by many authors as the hamartomatous lesion affecting the maxillofacial region more commonly.

The three common variants of fibrous dyplasia like polyostotic fibrous dysplasia craniofacial fibrous dysplasia and monostotic fibrous dysplasia are reviewed in detail in this article.

Keywords: Fibrous dysplasia, management, McCune Albright Syndrome, monostotic, polyostotic.

Introduction

Fibrous dysplasia (FD) is a development skeletal disorder characterized by replacement of normal bone by a benign fibrous connective tissue matrix which displays varying woven bone, mature bone or cementicles. However, the term is largely descriptive, nosologically limited, and diagnostically nonspecific and describes only a process. Moreover, all variations demonstrate similar presentation and attempt to classify such lesions by relying solely on their microscopic appearance is difficult.Yet another difficulty with these lesions is that although the biologic range of these diseases spans from developmental, reactive, and neoplastic etiologies, they all behave in a benign fashion. FD accounts for 2.4% of all tumourous and tumour like lesions [1]

Lichtenstein introduced the term fibrous dysplasia, as a designation for multiple bone lesions. [2] FD is usually classified broadly as monostotic fibrous dysplasia and polyostotic fibrous dysplasia. [3,4] When the lesion affects one bone it is termed monostotic and when multiple bones are involved it is called polyostotic. The polyostotic type is less common than monostotic type, can present with only bone lesions or may be associated with cutaneous pigmentation i.e. Cafe-au-lait spots (Jaffe-Lichenstein syndrome) and disorders of endocrine system like pituitary adenomas, precocious puberty or hyperthyroidism. This severe form of fibrous dysplasia subsequently became known as the Albright triad or McCune-Albright syndrome [5,6]. About 10% of craniofacial bones are affected in monostotic fibrous dysplasia and 50%–100% of cases are affected in polyostotic fibrous dysplasia.[7,8,9] Etiopathogenesis and molecular biology.

The etiopathogenesis of FD remains largely unproven but different theories proposed that FD of bone was a developmental defect of the bone due to trauma, mal-development of mesenchymal tissue of congenital origin,

liver damage glandular dysfunctions and infections of fibrous dysplasia. It is also seen that FD becomes inactive or stabilized after the normal period of skeletal growth come to an end thereby contradicting neither the familial nor the hereditary etiology.

In the present years, FD is considered a sporadic condition (not believed to be hereditary) that results from a post zygotic mutation in the GNASI gene (Guanine Nucleotide binding protein, α – stimulating activity polypeptide 1 gene) [10,11,12]. This gene encodes a G protein that stimulates the production of c-AMP. The mutations results in a continuous activation of the G-protein leading to over production of c-AMP in affected tissues [13,14]. The GNAS mutation can be detected in circulating mononuclear cells, bone cells, melanocytes and endocrine cells. The probability of detection is proportional to the number of mutated cells and the severity of the disease. Looking for the mutation may be useful to establish the diagnosis while avoiding a bone biopsy, or to confirm a pathological result because the pathological examination is sometimes challenging.

Monostotic Fibrous Dysplasia

Approximately, 80-85% of fibrous dysplasias are monostotic affecting frequently the ribs, femur, tibia, craniofacial bones and humerus in the descending order of frequency. This form may present in the age range of 10-70 years with mean age of 27 years. This is seen due to the hyperfunction of the endocrine organs, like hyperthyroidism, growth hormone, precocious puberty and cortisol overproduction [15]. There is an increased proliferation of melanocytes resulting in large café-au-lait spots. Also, cAMP is thought to have an effect on the differentiation of osteoblasts leading to fibrous dysplasia. The condition presumably depends on the time during fetal or post natal life where the mutation of GNASI occurs. Multiple bone lesions, endocrine disturbances, and cutaneous pigmentation, would result when the mutated cell carry the mutation and express the mutated gene. The mutated cell will disperse and participate in the formation of the skeleton resulting in multiple bone lesions of fibrous dysplasia if the mutation occurs during thelater period, and confined to one site affecting a single bone in the postnatal life.

Clinical Features

In FD replacement of all the components of cancellous bone by fibrous tissue is seen due to a change in normal bone metabolism. Hence, the appearance of numerous short, irregularly shaped trabeculae of woven bone is seen, which is responsible for the histological appearance and the internal pattern of the radiographs.

FD accounts for 2.4% of all tumors and tumor-like lesions. It most frequently occurs in the metaphyseal and diaphyseal areas of the long bones, the shoulder bones, the bones of the pelvic girdle and those of the jaw and the skull [16].

FD of the jaws is basically a disease of children, adolescents, and young adults, that tends to stabilize and essentially stops growing as skeletal maturity is reached. It affects both sexes equally, except for McCune-Albright syndrome, which is seen affecting females most commonly. It is common in the maxilla slightly more often than the mandible. The molar, premolar and canine areas, the ramus, and the symphysis are the most frequent sites in the mandible. In the maxilla, fibrous dysplasia might extend into the floor of the orbit, zygomatic process, and backward towards the back of the skull, as well as through the sinus. Patients with jaw involvement first may complain of unilateral painless facial swelling or an enlarging deformity of the alveolar process. The lesion is a low plateau, nodular, or dome-shaped and is firm, smoothly contoured, and covered with normal mucosa. Teeth in the region remain firm and are not displaced.The

main symptoms are swelling and pain and the associated symptoms are neurologic symptoms such as anosmia (loss of the sense of smell), sinusitis, deafness, blindness and pathologic fractures. Oral contraceptives may make the lesion active in pregnant females usually after a surgical intervention in young patients.

MFD is less severe when compared to polyostotic type in terms of the bone deformity. There is swelling of the labial or the buccal plate and seldom the lingual aspect with tipping or displacement of teeth due to progressive expansile nature of the lesion and tenderness.

Polyostotic Fibrous Dysplasia (PFD)

Approximately 20-30% of FD is polyostotic. This frequently involves the skull and facial bones, pelvic bones, spine and shoulder girdle. The dysplasia may be unilateral or bilateral and it may affect several bones of the limbs with or without axial skeletal involvement. Two-thirds of the patients are symptomatic before the age of 10 years. Pain associated with a limp, spontaneous fracture or both is seen commonly in the weight-bearing bones. The curvature of the femoral neck and proximal shaft of the femur markedly increase causing a Shepherd's crook deformity which is a characteristic sign of the disease.

The 2 of variants ofPFD are Jaffe's type and McCune Albright syndrome (MAS).

Jaffe's type involves a variable number of bones, accompanied by pigmented lesions of the skin or café-au-lait spots and this pigmentation are well defined, unilateral tan macules trunks and thighs. This is in contrast to café-au-laitspots of neurofibromatosis which have smooth borders.

MAS is involves nearly all bones in the skeleton and accompanied by pigmented lesions of the skin, and in addition endocrine disturbances, like sexual precocity and other endocrine manifestation of the syndrome.[5,6]

Craniofacial Fibrous Dysplasia

The designation of craniofacial fibrous dysplasia is appropriate for lesions which often involve multiple facial bones like the frontal, sphenoid, maxillary and ethmoidal bones zygoma, are not strictly monostotic. The pattern of the disease occurs in 10-25% of monostotic forms and in 50% with polyostotic form. It also occurs in an isolated form. The occipital and temporal bones are less commonly affected. The involvement of these bones causes esthetic and functional disorders. [16,17]

Radiologic Features

Location: Fibrous dysplasia involves the maxilla almost twice as often as the mandible and occurs more frequently in the posterior aspect. Lesions more commonly are unilateral except for very rare extensive lesions of the maxillofacial region that are bilateral.

Periphery: Most commonly the boundaries of the lesion are ill-defined, but sometimescan be well-defined especially in young lesions.

Internal structure: The variation and bone density is more pronounced in the mandible and more homogeneous than in the maxilla and more radiopaque in the maxilla and the base of the skull. The ground-glass appearance is seen due to abnormal shorter, thinner, irregularly shaped trabeculae more numerous than the normal trabeculae. The other common radiographic appearance is the peau d' orange, or a wispy arrangement (cotton wool), or an amorphous, dense patternor a "smoke screen appearance". There is a coarse mottling with irregular radiolucent and radiopaque areas, which involve the tissues surrounding the teeth although the teeth themselves are unchanged. The other appearance is an ill-defined radiolucent area with a few faint trabeculae. Also, a chalky type appearance composed of dense, amorphous material has been described.Occasionally bone cavities that are analogous to

simple bone cystsmay occur in mature lesions of fibrous dysplasia.

Effects on surrounding structures: The expansion of the involved bone may cause thinning of the outer cortex or involvement of the sinus, nasal cavity, internal ear canal stenosis, displacement of the eyeball.Often the periodontal ligament space becomes very narrow and the lamina dura surrounding the teeth disappears. Displacement of the teeth or interference with the normal eruption, root resorption, and displacement of the inferior alveolar nerve canal in a superior direction is also observed.

Radiologically distinct forms of fibrous dysplasia occurring in the maxillofacial area [18],

1. The pagetoid type: Lesions expands the skull outwards, thinning, inflating, buckling and displacing the outer horizontal table of the frontal bone. The sphenoid bone is often affected in the same way. The inner table is substantially expanded and denserand there are numerous points at which it becomes loosened from the bone surrounding it.
2. The sclerotic type: This type consisted of a massive thickening of the entire base of the skull including the orbital region. This sclerotic type of fibrous dysplasia appears as a horizontal plate located along the lower rim of the orbital cavity. It is poorly delineated, has high calcium content and can be up to 2 cm in width.
3. The cyst-like type: They are oval or rosette-shaped lesions of 2-5 cms in diameter, surrounded by a thin sclerotic border. In cases where there is a single core, the lesion is similar in appearance to eosinophilic granuloma and when multifocal, the lesion resembled a Hand- Schuller-Christian condition. The radiolucent, unilocular, cyst like variety are more common in the mandible,

Other advanced imaging modalities which can be highly useful to delineate the exact extent of the lesion is the CT, 99mTc radionuclide imaging fibrous was used, to delineate the boundaries of the lesion.Bone scans with 99mTc can detect the bone sites affected by FD because of increased uptake. C.T. could establish the extent as well as the involvement of orbit.

Histological features

There are considerable microscopic variation showing proliferating fibroblasts in a compact stroma of interlacing collagen fibers and irregular bone trabeculae or a c-shaped or Chinese character shaped trabeculae scattered throughout the lesion with no definite pattern of arrangement is seen. Three site-specific histopathological patterns are identified in fibrous dysplasia as follows [19];

a. Chinese writing type; associated with the axial and appendicular skeleton.

b. Sclerotic/ pagetoid type, associated with cranial bones.

c. Sclerotic/ hypercellular type; associated with maxilla and mandible.

The jaw lesions mature with time and may show lamellar bone. The bone in FD has an abnormal component of bone matrix components.

Imaging

Diagnosis is usually easy in PFD and MAS because of the grossly hemimelic distribution of typical bone lesions. In contrast, the diagnosis may be difficult in monostotic forms. The most common differential diagnoses are Paget's disease of bone, cherubism, meningioma, angioma and osteofibrous dysplasia. CT can be helpful in some instances to establish the diagnosis. The most important roles of CT are to give information on the size of the bone lesion and on cortical erosions that may not be visible on plain radiographs and to detect fissures. It is also useful to assess potential nerve compression, particularly for optic canal narrowing. Density reading scan be used to distinguish FD from other conditions such as osteomyelitis, Langerhans granulomatosis and some

malignancies in which bone density is significantly lower. Incontrast, CT cannot rule out other cystic conditions.The fibrous tissue is responsible for the low-intensity signals observed on MRI T1-weighted and spin-echo sequences. Variable intensity signals, especially high-intensitysignals in T2-weighted sequences, are a consequence of the heterogeneous histologyof FD and of metabolically active lesions.39 Non-specific liquid intensity signals areencountered in cases of cystic FD lesions.

Bone biopsy

A bone biopsy is necessary whenever there is a sizeable doubt on the diagnosis with imaging.The risk of the fracture induced by the procedure, however, needs to be considered.

Laboratory studies

Laboratory studies have shown that the cells surrounding the woven bone produced a matrix containing high amounts of osteopontin and bone sialoprotein (pro-adhesion protein); the reverse of which occurs in normal lamellar bone.

Serum levels of calcium and phosphorus are not usually altered in FD unless there is a related endocrinopathy. There may be a slight elevation of serum alkaline phosphatase depending on the extent of bone involvement. Urinary hydroxyl proline and urinary N-telopeptide may be elevated, reflecting bone collagen turnover.

Differential diagnosis

As proposed by Wood and Goaz [20], the differential diagnosis for FD should include many aspects. In the early stages, when FD can present as a solitary radiolucency with ragged and poorly defined borders, it should be differentiated from chronic osteitis where pain and other signs of inflammation, the presence of odontogenic cause like a carious tooth with pulp involvement or a nonvital tooth which is tender on percussion is present.

While chronic osteomyelitis confirms presence of signs of infection including inflammation, tenderness, pain, swelling, intraoral and extraoral sinus tracts, regional lymphadenopathy, fever, leukocytosis and increased sedimentation rate. In osteomyelitis of the jaws, the new bone is laid down on the surface of the outer cortex whereas in fibrous dysplasia, there is expansion of the internal aspect of bone, displacing and thinning the outer cortex. In Osteomyelitis, radiographs show irregular cortices of bone at least at one point along the expanded region. This is almost never so in FD, in which the expanded periphery, although thinned, appears smoothly contoured and basically uniform. The identification of sequestra aids in the identification of osteomyelitis.

Peripheral and central squamous cell carcinoma occurs in elderly with distinctive clinical features like mass in the oral cavity, pain, paresthesia and rapidly enlarging swelling of the jaws. Moreover, the lesions of FD are usually located deep within the jaw rather than superficially in the alveolus. Metastatic tumor of the jaws has a positive signs of pain and numbness as common complaints. A history of symptoms of or treatment for a primary tumor elsewhere is usually elicited during the history. Malignant minor salivary gland tumors are a rare entity and present with a mass in the oral cavity. Osteogenic sarcoma is a osteolytic type of tumor which may produce a similar pattern occurring in the older age group and shows malignant features. Chondrosarcoma usually occurs in older age group and causes a painful swelling of the jaws [21].

During the mixed radiolucent-radiopaque stage, FD should be differentiated from chronic osteomyelitis, osteoradionecrosis, and periapical cement osseous dysplasia where the distribution of bone pattern is often

bilateral, with an epicenter in the periapical region more commonly seen in the older age group. Focal cement osseous dysplasia occurs in older individuals. Paget's diseases may produce a similar pattern and may cause expansion, but it occurs in an older age group where the whole mandible is involved, unlike the unilateral tendency of fibrous dysplasia.

The differentiation of fibrous dysplasia from cemeto-ossifying fibroma (COF) can be very difficult. The differences recognized are:

- Shape: The lesions of the COF are predominantly rounded, whereas those of FD are more rectangular.
- Jaw expansion: Jaw expansion caused by COF is usually nodular or dome shaped, whereas that of FD is usually of the elongated fusiform type. The expansion of the jaws associated with COF is more concentric about a definite epicenter, but fibrous dysplasia enlarges the bone while distorting the overall shape to a smaller degree; in other words, the expanded bone still resembles normal morphology.
- Predominant Jaw: Approximately 70% of COF occur in the mandible whereas FD has slight predilection for maxilla.
- Predominant age: The age range of COF is 7 to 58 years and average is 26.4 years, whereas FD is found in patients below 20 years.
- Margins: The boundaries of a COF lesion usually are better defined, and these lesions occasionally have a soft tissue capsule and cortex, whereas fibrous dysplasia usually blends in with surrounding bone.
- Internal pattern: The internal structure of fibrous dysplasia lesions may be more homogeneous and show less variation.
- Effect on surrounding structures: Both types of lesions can displace teeth, but COF displaces from a specific point or epicenter. Fibrous dysplasia rarely resorbs teeth. Fibrous dysplasia usually displaces the lateral wall of the maxilla into the maxillary antrum, maintaining the outer shape of the wall, whereas an ossifying fibroma has a more convex shape as it extends into the maxillary antrum. Also, fibrous dysplasia alters the bone around the teeth without displacing them from an obvious epicenter of a concentrically growing benign tumor. The importance of this differentiation lies in the treatment, which is resection for an ossifying fibroma and observation for fibrous dysplasia.
- Histologic: Tiny calcified spherules may be seen rarely but are never numerous. In contrast to ossifying fibroma and cemento-osseous dysplasia, FD typically demonstrates a rather monotonous pattern throughout the lesion. Osteogenic sarcoma- the radiographic pattern of OS is more disorderly than that seen with fibrous dysplasia. Generally, one of the radiographic features of osteogenic sarcoma is apparently either a sunburst appearance, cumulus cloud appearance, Codman triangle, asymmetric, band-like widening of the periodontal ligament space, and an onionskin appearance of redundancy of the cortical plate.

Osteoblastic metastatic carcinoma does not show a monotonous pattern like that of FD. A history of symptoms of or treatment for a primary tumor elsewhere is usually elicited during the history. Chondroma and chrondrosarcoma affects the older individuals and produces pain and shows a distorted image once the cortex is invaded. Hyperparathyroidism can alter the bone in a similar fashion but these diseases are polyostotic and bilateral and unlike fibrous dysplasia, do not cause bone expansion.

With spontaneous healing of a simple bone cyst, the radiographic and histologicalappearance of the new one may be very similar to that of fibrous dysplasia.

Complications

The occurrence of a fracture may reveal the disease or appear as the complication of known FD. Fractures of all involved bones can be observed. Stress fractures or fissures are common causes of bone pain. They can heal or become a complete fracture. Fissures may be missed on plain radiographs, so the occurrence of bone pain in a previously asymptomatic affected bone must lead one to consider using computer tomography (CT). In patients with severe FD, the peak fracture rate is between 6 and 10 years of age,although there is still a sustained incidence of fracture in adulthood.

Deformities are observed in craniofacial bones and long bones. They may be responsible for significant functional disability and aesthetic discomfort. Patients with severe PFD often have spinal involvement, which is associated with scoliosis in 40% of cases.Unlike idiopathic scoliosis, curve progression after the end of growth is frequent. It is unknown whether bracing will prevent progression, but surgical fixation seems to be as effective as for idiopathic scoliosis.

Neurological complications result from nerve compression due to expansion of FD lesions. Consequently, patients with spinal involvement may suffer from spinal cord compression. Deafness has also been described in cranial FD. The most common neurological complication is blindness related to optic nerve compression. The optic canal diameter of patients with craniofacial FD must be evaluated using CT. Patients with optic canal narrowing must be followed-up carefully, with frequent measurement of their visual acuity and visual field. The long-term prognosis, however, remains good because, in a series of severely affected patients followed in the long term, 86% retained normal vision and only 5% had loss of vision, with no age influence. As such, prophylactic optic nerve decompression is contra-indicated, and surgery should only be discussed in cases of visual impairment. The only risk factor predicting poor visual outcome is growth hormone excess in MAS.

Malignancies are rare in FD but may be observed both in monostotic and polyostotic forms. In large series, the frequency of malignant transformation varies between 0.5% and 4%, although this incidence may have been overestimated because many of the patients had received radiotherapy. Radiotherapy is contra-indicated as it appears to be the main risk factor for malignant transformation [22]. The most common histology is osteosarcoma, followed by fibrosarcoma and chondrosarcoma. Patients have rapidly developing symptoms such as pain and swelling. The characteristic finding on radiograph is expanding lesion in soft tissues, through the cortex. When the diagnosis is suspected, CT and MRI are the best non-invasive techniques to suggest a provisional diagnosis of malignant transformation of FD.

Management

Management of fibrous dysplasia of the jaws may present a major problem. The smaller lesions, particularly in the mandible may be surgically resected, the large and diffuse lesions, particularly those of maxilla, may require surgical recontouring for correction of cosmetic or functional deformity.

Radiation is contraindicated in FD due to malignant and sarcomatous changes [22]. The hormonal changes especially in females due to pregnancy or the use of oral contraceptives may stimulate the growth or result in the development of lesions with in the area of fibrous dysplasia, such as aneurysmal bone cyst or giants cell granulomas. From a prognostic view point, FD does not constitute a direct threat to life, and monostotic masses require no treatment, providing they are causing no functional disturbance.

Non surgicalpharmacological treatment modalities have been tried with varying success.

Bisphosphonates

Bisphosphonates drugs have been used for fibrous dysplasia with good success rate [23]. The drugs include pamidronate (180 mg every 6 months), alendronate (10 mg/day, as an adjuvant to pamidronate) risedronate (30 mg/day for two consecutive months every 6 months) and Zoledronic acid [24]. Biphosphonates are a family of drugs and analogues of pyrophosphate (P–O–P) in which the oxygen atom between the two P-atoms is replaced by a carbon atom (P–C–P) opening up the possibility of attaching side chains, i.e. hydroxyl, alkyl or amino groups. The biphosphonates control bone erosion bythe inhibition of osteoclast action. Due to its structure they present high affinity for the hydroxyapatite of resorbed bone and remain tied to it for a longperiod (several months or even years). Osteoclasts resorbingthe area of bone covered by the drug incorporate the agent into the cytoplasm where it inhibits acid phosphatase secretion thereby arresting bone resorption. Pamidronate containing a basic nitrogen atom in its alkyl side chain represents a second-generation drug, characterized by increasedpotency of inhibition of bone resorption and good tolerance. Latest therapies using drugs like denosumab, which is a inhibitor of Receptor activator of nuclear factor kappa-B ligand (RANKL), also known as tumor necrosis factor ligand superfamily member 11 (TNFSF11) is seen to be effective in the treatment of FD-related pain or reduction in growth remains to be determined [25].

Supplements

Significant mineralization defects are common within the dysplastic bone vitamin D deficiency is observed frequently and hyperparathyroidism-related changes correlated with serum PTH may be observed on histological examination.These findings support the use of calcium and vitamin D supplements in the subset of FD patients with deficiency, although the use of such supplementshave not shown to reverse the histological appearance of hyperparathyroidism-related changes.These mineralization abnormalities may also relate to the renal phosphate wasting observed in many patients with PFD. Therefore, supplementation with oral phosphorus (associated with calcitriol) appears logical in patients with hyperphosphaturia, although it has not yet been proven efficacious in clinical trials. However, these supplements are useful patients with osteomalacia and hyperparathyroid changes related FD in reducing the severity of FD.

Fibrous dysplasia has often been confused with osseous lesions of hyperparathyroidism indicating similar histopathologic features. Parathyroid hormone (PTH) is a peptide hormone produced in the parathyroid gland which acts directly on bone to increase bone resorption and mobilize calcium to control the concentration of calcium in the extracellular fluid; thisfunction is effected through activation of a mechanism that transfers calcium from bone and from glomerularfiltrate to the extracellular fluid compartment. Parathyroid hormone-related peptide (PTHrP) is elevated and concomitant increase in PTHrP, mRNA expression are reported in cultured osteoblasts from thepatients with fibrous dysplasia in the McCune Albright syndrome. Fibrous dysplasia associate with a particular endocrinological disorder of bone maturation. Calcitonin could be classically considered to be aphysiologic antagonist of PTH and PTHrP; it lowers the plasma calcium concentration by inhibiting bone resorption, with a stimulation of the renal calcium excretion. Moreover, Calcitonin has a role in protecting the skeleton in times of calcium stress, such as pregnancy and lactation. Calcitonin and 1,25-dihydroxy vitamin D3

have been used for the treatment of fibrous dysplasia associated with McCune Albright syndrome.

Gene therapy would imply silencing the gain-of-function mutated gene. Further research is needed in this area.

Surgical management

Surgery is indicated for the correction of deformities, the prevention of fracture in patients with threatening osteolytic lesions, the management of fractures, and transformation of lesions into either benign (i.e. bone cyst) or malignant tumors. In PFD, the bones are often thin and weak, so the choice of technique has to be considered carefully before the intervention. With most lesions, growth is complete at skeletal maturation; therefore orthodontic treatment and cosmetic surgery may be delayed until this time. A corrective recontouring osteotomy should be performed when necessary. If spontaneous fractures occur, reaction and immediate reconstruction with material from the ribs or pelvic girdle is advisable. Surgical intervention should only take place once the bone concerned has completed their growth period.

The prevalence of regrowth after surgical reduction is difficult to determine, but it has been estimated that around 25% to 50% of the patients show regrowth after surgical shave down of the lesion. This is especially seen in of recurrence in cases of fibrous dysplasia.

Therefore, the recommended treatment for FD includes:

1. Early and aggressive screening for efficient management of endocrinopathies (particularly growth hormone excess).
2. A detailed history, clinical examination and radiographic analysis are very essential especially in lesions where a confirmatory biopsy is not possible.
3. Surgical treatment is delayed till until the skeletal maturity in lesions which are quiescent.
4. However, if there are symptoms or rapid changes in the lesion then a surgical contouring or resection may be carried out prior to skeletal maturity, though a high risk of recurrence is seen.
5. Immediate surgical intervention and evaluation is done for active disease (rapid growth, new onset of pain or paresthesia, visual or hearing changes).
6. The use of bisphosphonates may be considered as an adjuvant for refractory pain at the site of the lesion.
7. A multi-disciplinary approach is vital for the successful management of patients with FD, particularly PFD and MAS [26].

Summary and Conclusion

In summary, FD of the maxillofacial region comprises a diverse group of diseases which manifests themselves microscopically in almost similar fashion and therefore is indistinguishable from each on morphological basis alone. There is replacement of normal bone by fibroblasts, collagen fibers and mineralized tissues. FD comprises of developmental, reactive and benign neoplastic forms. A number of other disease processes involving the maxillofacial region may be clinically, radiologically and / or histologically confused with the "conventional" fibro-osseous lesion of the jaws. These include some types of chronic osteomyelitis, periosteitis, Paget's disease, hyperparathyroidism, osteoblastoma, and low grade osteosarcoma. Separation of these various types of lesions can usually be accomplished by thorough evaluationof the clinical, radiologic and histologic features, although in some cases this may be very difficult. Thus, the definitive diagnosis and management of FD usually requires close communication between clinician and pathologist, because clinical, radiographic, operative, and microscopic features must all be considered together.

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