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Review Article

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Updated systematic review: ossifying fibromas of the nose and paranasal sinuses

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ABSTRACT

Ossifying fibroma is one of the benign fibro-osseous lesions (BFOL), which is a relatively rare group of diseases of the bone with diverse pathologies and involving different parts of the body and mostly craniofacial bones. BFOLs share a common histopathologic feature of replacing the normal bony tissue structure with a distorted fibrous and mal-arranged bony structure. Although, ossifying fibroma is pathologically benign, but it's presentation can vary from an incidental radiologic finding to a devastating facial mass with cosmetic and functional disabilities. And the management is according to the site involved and the symptomatology. The purpose of this systematic review is to collect the available recent data regarding ossifying fibromas involving nose and paranasal sinuses, to have a better understanding of the disease, and its management. This review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement and PRISMA checklist. The U.S. National Library of Medicine (Pubmed) database was searched. Abstracts were screened by two authors and the ones that met the inclusion and exclusion criteria were included. Additional articles were retrieved from the citations in the previously found articles. A total of 156 distinct cases were found. Demographic data, clinical presentation, radiological and pathological findings and the management approaches were reviewed. Review of the recent data highlighted new diagnostic, imaging and management strategies that should be considers in the approach to patients with ossifying fibroma.

Keywords: Ossifying fibromya, Benign fibro-osseous lesions, Cemintifying fibroma, Psammomatoid ossifying fibroma, Psammo-osteoid fibroma, Sinonasal masses

INTRODUCTION

Benign fibro-osseous lesions (BFOL) are a diverse group of diseases involving various bones of the body, but mainly the craniofacial bones and the mandible. They are characterized by replacement of the normal bony architecture by a haphazard benign fibre-osseous structure. Despite the recent advancement in the molecular analysis BFOL; are still difficult to differentiate due to their pathological similarities. Numerous trials for classification of BFOLs have been proposed and some of them have further classified ossifying fibromas into different groups.

In 1985, Charles Waldron classified OF into two groups according to their origin. Those arising from periodical ligament and classified as ossifying and cementifying fibroma and those with uncertain or detectable relationship to the periodontal ligament then classified as juvenile active ossifying fibroma and other aggressive ossifying/cementifying fibromas.[1] Following that, in 1990, Slootweg and Hellmuth Muller, and in 2001

Brannon and Fowler, categorized OFs into two categories; ossifying fibroma and juvenile ossifying fibroma, based on their histopathological features.[2,3] Speight in 2006, proposed a three category classification of conventional ossifying fibroma, juvenile trabecular ossifying fibroma and juvenile psammomatoid ossifying fibroma.[4]

The most detailed classification was proposed by Eversole et al in 2008, in which not only the histopathological features but also the clinical and radiological findings were considered. They divided the OFs into ossifying fibroma, hyperparathyroidism jaw lesion syndrome and juvenile ossifying fibroma. The latter was further divided into trabecular type, psammomatoid type, gigantiform cementomas.[5]

METHODS

This review was performed according to the preferred reporting items for systematic reviews and meta-analyses (PRISMA) statement and PRISMA checklist. The U.S. National Library of Medicine (Pubmed) database was searched. The search was conducted on the articles of the last 10 years starting from 25 October 2018. Including search terms were: ossifying fibroma, cementifying fibroma, cemento-ossifying fibroma, desmo-osteoblastoma, psammo-osteoid fibroma, psammonmatoid ossifying fibroma, juvenile ossifying fibroma, juvenile aggressive ossifying fibroma and juvenile active ossifying fibroma. And a language restriction to English papers was applied.

All age groups and both genders were included. This study included ossifying fibroma in the nose and paranasal sinuses and studies with disease involving other sites of the body were excluded. Studies in non-English language, animal studies, duplicated studies and reviews and studies with no histopathologic confirmation of ossifying fibroma were excluded. The abstracts reviewed by the two authors (J.I and M.A.) independently for inclusion and inter-reviewer agreement was assessed using Cohen' Kappa scores. Disagreement resulted in exclusion of the articles.

Statistical analysis was conducted by SPSS software. Abstracts were screened by two authors and the ones that met the inclusion and exclusion criteria were included. Additional articles were retrieved from the citations in the previously found articles.

RESULTS

A total of 156 distinct cases were identified.[6-28] Demographic data, clinical presentation, radiological and pathological findings and the management approaches were reviewed. The PRISMA flow chart for article selection procedure is shown in Figure 1. Cohen's kappa for inter-reviewer agreement was 0.75 that is considered excellent.

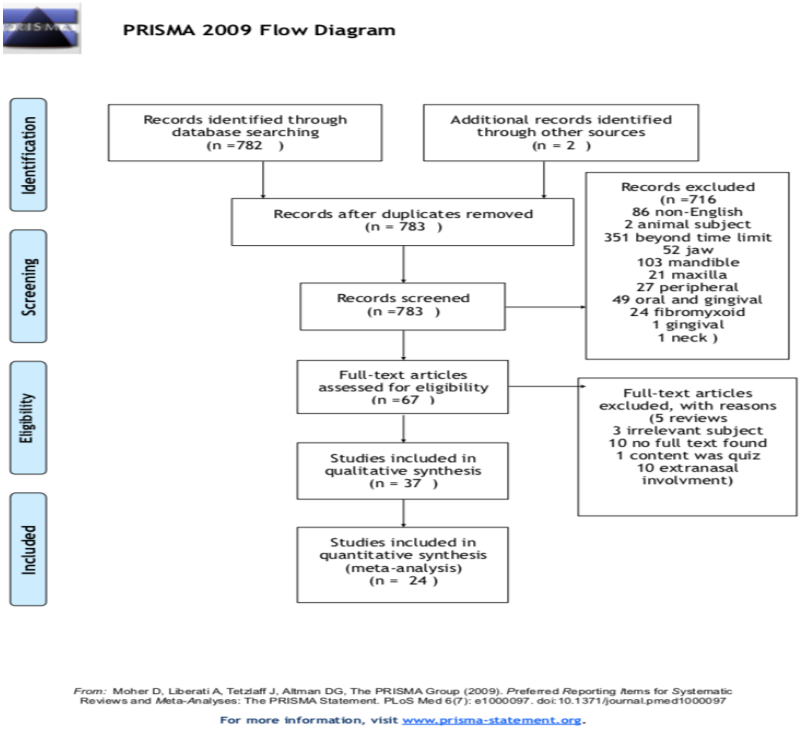

Figure 1: Flow diagram of article selection.

Age range was 2 months to 68 years. Mean age of presentation in both sexes was 23.23 years and standard deviation of 16 years old. Half of the affected cases were presenting between the ages of 10-30 years. Males and females were equally affected. However, females were having more frequent presentation in elder ages, with 28% above 40 years of age while males only having 8% of cases above this age. 50% of male cases are below 20 years while only 30% of females were presenting in the same age group Figure 2.

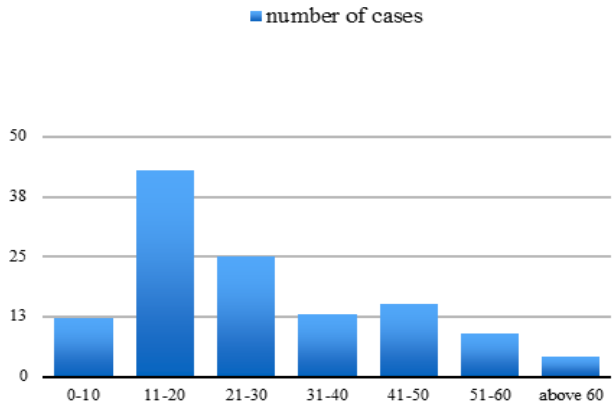

Figure 2: Age distribution of cases.

The patients presented with symptoms related to the expansion of the mass, namely proptosis, nasal obstruction, headache, facial masses and deformities, visual disturbances related to the displacement of the orbit, epiphora and hyposmia. Other symptoms included rhinorrhea, recurrent sinusitis and epistaxis (Table 1).

Table 1: The presenting symptoms of the patients.

Symptoms at presentation	No. of patients N (%)
Proptosis	40 (31)
Nasal obstruction	39 (30)
Headache	29 (23)
Visual disturbance	19 (15)
Facial mass and deformity	8 (6)
Rhinorrhea	7 (5)

Table 2: CT based site of involvement.

Site involved	No. of patients N (%)
Ethmoid sinus	72 (56)
Sphenoid sinus	38 (29)
Orbital wall	38 (29)
Maxillary sinus	27 (21)
Frontal sinus	19 (14)

Review of the CT findings showed that most of the lesions included the skull base with the mass seen in the ethmoid and sphenoid sinuses (Table 2).

Only 61 of the total cases had reported histopathologic classification, with differentiating juvenile from conventional type OFs. Conventional type showed female predominance with male to female ratio of 1:2 and mean age presentation of 36.2 years and standard deviation of 18.8. While juvenile type had male predominance with male to female ratio of 2:1 and a much lower mean age presentation of 16.1 years and standard deviation of 9 .

Of all reported cases 11% (18 patients) had recurrence with mean age of 20 years. Histopathological classification was only available for 6 of these cases and all of them were juvenile type OFs.[6-8,11,20]

DISCUSSION

Etiology

Ossifying fibroma is a disease with benign growth of a mal-arranged architecture of bone and fibrous tissue. Some authors suggested a genetic component for OFs, but very few articles have described the genetic component of OFs. Dal et al was first to describe deletions in 2q31-32 q35-36 that was detected in a single case of OF.[29]

In 1995, Sawyer et al described a consistent chromosomal defect at bands Xq26 and 2q33 in three patients in which two of them had identical reciprocal translocation and the other one had an interstitial insertion.[30]

However, it is difficult to conclude a constant defect with this small number of patients and no other studies have found similar results or supported their findings. A more recent study by Tabareau et al showed another and yet different genetic defect. This study was larger than the previously mentioned studies and it included 30 cases of OFs. Tabareau et al found a modified chromosome 12 with MDM2 and RASAL1 amplification in 69% cases of juvenile OFs, that was statistically significant (p value 0.0001) in differentiating juvenile OFs from fibrous dysplasia and non-juvenile OFs. They speculated that the presence of this genetic defect can be associated with the aggressiveness of the disease and predict higher recurrence rate. But the small number of cases and the presence of great confounding factors and more importantly the complete excision with no residual makes it difficult to make a firm conclusion.[10]

In another study Tabareau et al studied 91 cases with BFOL and confirmed the presence of GNAS mutation to be specific to fibrous dysplasia among all BFOLs. Despite the low sensitivity, the presence of this genetic mutation can be useful to rule out OFs.[12]

The form of the 4th Edition of the World Health Organization Classification of Head and Neck Tumour recognized CDC73 (previously known as HRPT2) to be the genetic alteration in OFs.[31]

Radiologic appearance

Radiological imaging for OFs should be performed to determine the extent of the disease and help in surgical planning. CT imaging is usually the first imaging used and can give a very precise anatomical localization in regard to bony boundaries. In early stages, the lesions can be radiolucent since the osseous component is still not calcified. But later, with calcifications of the osseous components, the lesions start to be more radiopaque. CT finding of a rim of well demarcated dense bony shell can suggest OFs in opposite to the other BFOLs especially fibrous dysplasia that usually has poorly demarcated edges. This is important to know, since OFs usually have a more aggressive nature than fibrous dysplasia and a more aggressive surgical resection should be attempted.[32]

MRI helps to determine soft tissue invasion by the lesion. OFs typically show an intermediate to low signal intensity on T1-weighted images with contrast enhancement and mixed low and high signal density on T2-weighted images.[33] Reiko et al was the first to describe a nuclear imaging with [18F] FDG and [11C] Met uptake in a case with JPOF, which was suspected to be due to increased protein metabolism within the lesion. [11C] Met PET/CT was suggested to use for detecting residual lesions after surgical removal.[27] Zhang et al reported a similar finding of 99mTc-MDP uptake in a whole body bone scan.[34]

Management

Ossifying fibromas are not always symptomatic and in asymptomatic cases a "wait and scan" strategy can be used.[23] The recommended management for the symptomatic ossifying fibromas is the complete excision and recurrence with partial excision was reported to be as high as 30-56%.[35] Due to the benign pathology of this disease it is generally accepted to preserve a shell over anatomically critical areas, mainly the carotid artery and optic nerve.[6]

Usually the OF is well demarcated and is like an egg with a shell. OFs can be shelled out or curetted from the surrounding bone with relative ease, which differentiate it from fibrous dysplasia that is poorly demarcated. During the resection of the mass bulk the bleeding will be more profuse, however when the bone shell of the tumor is exposed the bleeding will subside. Use of fibrin packing and bone wax can help to control bleeding.[6] In cases where excision of the bony shell is difficult, a diamond drill can be useful to grind the hard bone.[7] The exact approach to the lesion should be personalized to each case according to the location and the size of the disease. The mainstay of treatment of nose and paranasal sinuses and skull base is the endoscopic end nasal approach since it is less invasive, safe and has less complications and better cosmetic results than the open approaches. The advance of intra-operative navigation assisted techniques further enhanced the localization of the lesion and improves complete excision. One of the inherited deficiencies of intra-operative navigation is the inability to reflect the anatomical changes made by surgical intervention. Wise et al have suggested repeating the CT intra-operatively to give real-time update on the changes and have demonstrated significant improvement on the identification of the bony landmarks.[36] Wong et al have demonstrated the use of this technique on one case in which intra-operative CT-assisted in completion of lesion removal.[24]

Radiotherapy is traditionally considered to cause malignant transformation of OFs, and even with lack of evidence to support this theory, it is mostly abandoned. Scott et al presented a case of of treated with adjacent radiotherapy postoperatively and suggested radiotherapy to decrease recurrence. However the follow up for this patient was reported for only 7 months, which is not enough either for suggesting decreased recurrence or for rejecting the malignant transformation theory. More studies are needed to support this assumption.[13]

CONCLUSION

This review of recent literature on ossifying fibromas highlighted the updated trends in the diagnosis, imaging and management strategies. Since ossifying fibroma is an uncommon pathology most of the findings need yet to be supported with larger number of patients. However, knowing about the reported strategies and the results in the diagnosis and management that are used previously by other authors can guide the practitioner about his choices.

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