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CLINICAL REPORT

Ossifying Fibroma: A Case Report of an Unusual Presentation of Angiodysplastic Disease with Review of Literature

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Abstract Ossifying fibroma is a nonneoplastic developmental disease of osseous tissue seen rarely in association with Sturge–Weber syndrome. It is a lesion of unknown aetiology, uncertain pathogenesis, and diverse histopathology. The aim of this study is to report an unusual case of in a 11-year-old male of SWS. The rarity of the case and the fact that ossifying fibroma may be associated with Sturge–Weber syndrome propelled us to report it. Physical examination showed facial asymmetry (due to hemifacial swelling) without any tenderness, fluctuation, ocular pain, or ophthalmoplegia. Imaging studies revealed a solid mass involving the left maxilla and orbital floor. A conservative therapeutic approach to these lesions may be sufficient to relieve signs and symptoms effectively. Periodic follow-up is indicated to detect recurrences or malignant changes in the early stages.

Keywords Ossifying fibroma · Sturge–Weber syndrome · Maxilla · Orbit

Introduction

Benign fibro-osseous lesions occur when normal bone is replaced by cellular fibrous connective tissue and mineralized structures. One rare type of these lesions is the ossifying fibroma (OF). Clinically, OF usually presents as a painless, expansive, central mass that is discovered incidentally. However, some cases present with pain, paraesthesia, nasal obstruction, aesthetic deformity, or orbital impairment [1]. In Sturge–Weber syndrome (SWS), where there is cutaneous capillary angioma involving the maxillofacial distribution, there are a few reported cases of described as angiodysplasia, and "angiodysplastic syndrome" implies a vascular malformation that is associated with secondary changes including further vascular abnormalities, soft tissue and bone hypertrophy. There are very few reported cases with osteohypertrophy with ipsilateral oromaxillofacial osseous overgrowth [2]. Extensive search of peer reviewed literature in English language revealed only 02 cases reported in India in last two decades and to the best of our knowledge this is the third case report where OF occurred in the maxilla in patient with SWS (Tables 1, 2, 3, 4).

Case Report

A 11-year-old male, known dystonic Cerebral Palsy and Acute Bilirubin Encephalopathy with G6PD deficiency diagnosed at the age of 2 years, presented with complaints of gradually progressive Right hemifacial (leading to facial asymmetry) and orbital swelling, associated with outward displacement of Right eye and reddish discolouration over face (bilateral cheek) of 03-year duration (Figs. 1, 2). Examination findings of face and nose revealed hard immobile swelling right cheek, not fixed to the underlying bone or overlying skin, right inferior turbinate occluding the right nasal cavity and eye examination showed right optic disc atrophy. CT PNS revealed a large well circumscribed expansive lesion in the right maxillary sinus measuring 3.8 × 3.2 cm with central osseous component displacing the globe superiorly. Bony erosion of the floor of maxillary sinus and adjacent upper alveolus suggesting Juvenile ossifying fibroma (Figs. 3, 4), MRI Brain and Orbit showed

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Table 1 Cases of reported ossifying fibroma (OF) in various syndromes around the globe (2000–2021)

Sl no	References	Year of publication	Journal	No. of cases	Age/sex	Presentation	Associated syndromes	Management	Histopathology	Follow up	Recurrence
1	de Vasconcelos Macedo et al. [1]	June 2020	Journal of the Korean Association of Oral and Maxillofacial Surgeons	01	57/F	Painless swelling in the left maxilla with facial asymmetry	None	Surgical excision	Areas of mature bone and a lesion composed of cellular fibrous tissue rich in fibroblasts and collagen fibers, confirmed the diagnosis of OF	50 months	Not reported till date
2	Babaji et al. [3]	April 2013	Case reports in pediatrics	01	08/F	Osseous abnormalities in oral cavity on right side	Sturge Weber syndrome	Maxillectomy was advised for enlarging maxilla. But patient's parents were unwilling for the surgical resection	–	Was lost for follow up	–
3	Jendi et al. [5]	November 2019	Indian Journal of Otolaryngology and Head & Neck Surgery	01	45/F	Painless swelling in right upper jaw	Neurofibromatosis 01	Enucleation of the tumour along with extraction of first premolar, second premolar, second and third molars	Features of a benign fibro-osseous lesion consistent with ossifying fibroma	Not reported	Not reported

Table 1 (continued)

Sl no	References	Year of publication	Journal	No. of cases	Age/sex	Presentation	Associated syndromes	Management	Histopathology	Follow up	Recurrence
4	Lin et al. [6]	February 2006	American journal of neuroradiology	01	17 month/M	A 4-month history of rapidly enlarging left face and maxilla, with complaints of drooling, malocclusion, inability to eat solid foods, failure to thrive, and difficulty breathing at night	Sturge Weber syndrome	Extended maxillectomy with en bloc resection of the roof of the left hard palate, followed by reconstruction	Consistent with an ossifying fibroma	Followed up 08 months later with a CT scan	Follow-up CT examination showed a markedly expansile lesion, in the same distribution of the cutaneous capillary malformation, involving the remaining left frontal–temporal skull extending to the orbital rims and maxillary sinus walls that had a typical ground-glass appearance of a fibro-osseous lesion
5	Saiz-Pardo-Pinos et al. [12]	November 2004	Medicina Oral, Patologia Oral y Cirugia Bucal	01	08/M	Swelling at the level of the mandibular angles and prevented the correct eruption of the lower first molars	–	Surgical excision	Consistent with an ossifying fibroma	Not reported	–

Table 1 (continued)

Sl no	References	Year of publication	Journal	No. of cases	Age/sex	Presentation	Associated syndromes	Management	Histopathology	Follow up	Recurrence
6	Ozek et al. [17]	May 2002	Journal of Craniofacial Surgery	16	Age group ranging between 8–36 years	Most of the lesions in the current study presented primarily as a painless swelling, which was reflected by expansion of the affected bone as seen on radiographs	2 patients-McCune Albright syndrome	All patients underwent surgical excision	Revealed fibrous dysplasia in all patients	All patients were followed up 03 monthly	All maxillary lesions in the eight patients who were treated with curettage relapsed during the year after the operation hence underwent secondary curettage
					Sex-10F,6 M						No recurrence was observed in these patients in later follow up visits

Table 2 Cases reported around the globe in last decade (2010–2021)

Sl no	References	Year of publication	Journal	No. of cases	Age/sex	Presentation	Associated syndromes	Management	Histopathology	Follow up	Recurrence
1	de Vasconcelos Macedo et al. [1]	June 2020	Journal of the Korean Association of Oral and Maxillofacial Surgeons	01	57/F	Painless swelling in the left maxilla with facial asymmetry	None	Surgical excision	Areas of mature bone and a lesion composed of cellular fibrous tissue rich in fibroblasts and collagen fibers, confirmed the diagnosis of OF	50 months	Not reported till date
2	Babaji et al. [2]	April 2013	Case reports in pediatrics	01	08/F	Osseous abnormalities in oral cavity on right side	Sturge Weber syndrome	Maxillectomy was advised for enlarging maxilla. But patient's parents were unwilling for the surgical resection	–	Was lost for follow up	–
3	Jendi et al. [5]	November 2019	Indian Journal of Otolaryngology and Head & Neck Surgery	01	45/F	Painless swelling in right upper jaw	Neurofibromatosis 01	Enucleation of the tumour along with extraction of first premolar, second premolar, second and third molars	Features of a benign fibro-osseous lesion consistent with ossifying fibroma	Not reported	Not reported

Table 3 Cases reported in India in last decade (2010–2021)

Sl no	References	Year of publication	Journal	No. of cases	Age/sex	Presentation	Associated syndromes	Management	Histopathology	Follow up	Recurrence
1	Babaji et al. [2]	April 2013	Case reports in pediatrics	01	08/F	Osseous abnormalities in oral cavity on right side	Sturge Weber syndrome	Maxillectomy was advised for enlarging maxilla. But patient's parents were unwilling for the surgical resection	–	Was lost for follow up	–
2	Jendi et al. [5]	November 2019	Indian Journal of Otolaryngology and Head & Neck Surgery	01	45/F	Painless swelling in right upper jaw	Neurofibromatosis 01	Enucleation of the tumour along with extraction of first premolar, second premolar, second and third molars	Features of a benign fibro-osseous lesion consistent with ossifying fibroma	Not reported	Not reported

a large, rounded lesion isointense on T1 and hypointense on T2 in the right maxillary antrum expanding and eroding the maxillary bone, causing obstruction of the right nasal cavity measuring 4.1 × 3.9 cm. Near complete opacification of right frontal, ethmoid and sphenoid sinus noted. A clinico-radiological suspicion of Sturge Weber Syndrome was made.

Patient underwent Endoscopic debulking of tumour right maxilla and histopathology of tumour showed features of benign fibro osseous lesion favouring monostotic fibrous dysplasia over ossifying fibroma. Patient was followed up with CECT PNS after a month, showed well defined lobulated expansive lytic lesion (41 × 47 × 56) mm involving (R) maxillary sinus and (R) maxilla causing expansion of maxillary sinus with thinning and contour bulge of its walls, Superiorly—Lesion causing contour bulge and thinning of inferior orbital wall and displacement of (R) globe and inferior rectus muscle superolaterally, Inferior-Eroding the floor of maxillary sinus and upper alveolus, Anterior-causing contour bulge of anterior wall of maxillary sinus, Posterior-abutting the (R) lateral pterygoid muscle with preserved fat planes and medial-Extending into (R) nasal cavity and displacing nasal septum to left, causing complete occlusion of (R) choana. Patient NoK counselled regarding benign nature, severity and progression of disease and advised for watchful follow up every 03 monthly or aggravation of orbital/Upper Aerodigestive tract symptoms and need for fresh imaging on such occasions to look for extent and progression of lesion.

Discussion

SWS is an uncommon nonhereditary developmental condition with neurological and skin disorder, also known as Sturge–Weber disease, encephalotrigeminal angiomatosis, meningofacial angiomatosis, and Sturge–Weber–Dimitri syndrome. It is a congenital hamartomatous malformation affecting the eye, skin, and central nervous system, with characteristic venous angiomas of leptomeninges, face, jaws, and oral soft tissues. The presence of angiomas result in alteration of vascular dynamics causing perception of calcium deposition in cerebral cortex underlying the angioma. This can result in seizures, mental retardation, hemiplegia, or hemiparesis.

SWS can show "tramline" or gyriform calcifications involving the occipital and parietal lobes on CT, MRI scanning, or on radiographs [3]. Cutaneous angiomas are called as port wine stains, having unilateral distribution along dermatomes supplied by the ophthalmic and maxillary division of trigeminal nerve. Sometimes can be bilateral or can extend up to neck, limb, and other parts of the body. Port wine stains in childhood are classically faint, pink macules, tend to darken to red purple, may be isolated with well delineated border, or may be very diffuse. Large lesions are warm

Table 4 Cases of ossifying fibroma (OF) of maxilla in Sturge Weber Syndrome reported (2000–2021)

Sl no	References	Year of publication	Journal	No. of cases	Age/sex	Presentation	Associated syndromes	Management	Histopathology	Follow up	Recurrence
1	Babaji et al. [2]	April 2013	Case reports in pediatrics	01	08/F	Osseous abnormalities in oral cavity on right side	Sturge Weber syndrome	Maxillectomy was advised for enlarging maxilla. But patient's parents were unwilling for the surgical resection	–	Was lost for follow up	–
2	Lin et al. [6]	February 2006	American journal of neuroradiology	01	17 month/M	A 4-month history of rapidly enlarging left face and maxilla, with complaints of drooling, malocclusion, inability to eat solid foods, failure to thrive, and difficulty breathing at night	Sturge Weber syndrome	Extended maxillectomy with en bloc resection of the roof of the left hard palate, followed by reconstruction	Consistent with an ossifying fibroma	Followed up 08 months later with a CT scan	Follow-up CT examination showed a markedly expansile lesion, in the same distribution of the cutaneous capillary malformation, involving the remaining left frontal–temporal skull extending to the orbital rims and maxillary sinus walls that had a typical ground-glass appearance of a fibro-osseous lesion

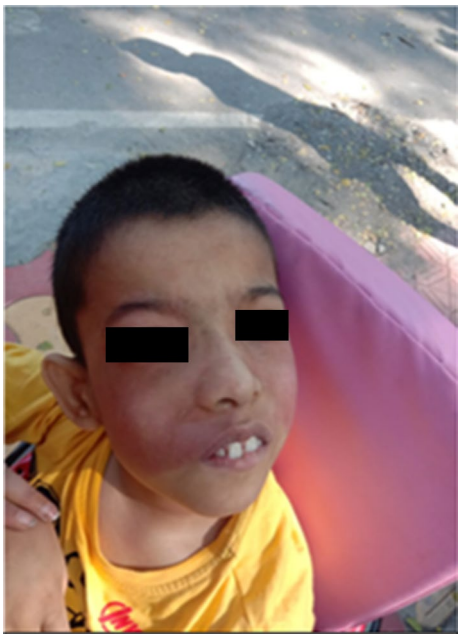

Fig. 1 Showing facial asymmetry over right face

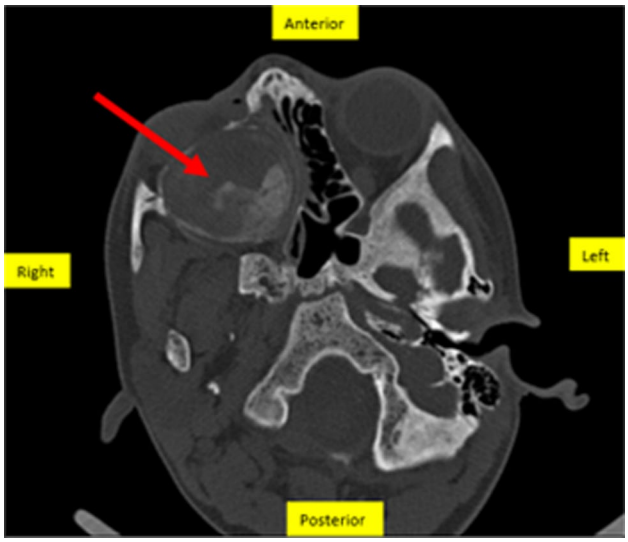

Fig. 3 Axial view of computed tomography scan showing lesion in right maxilla

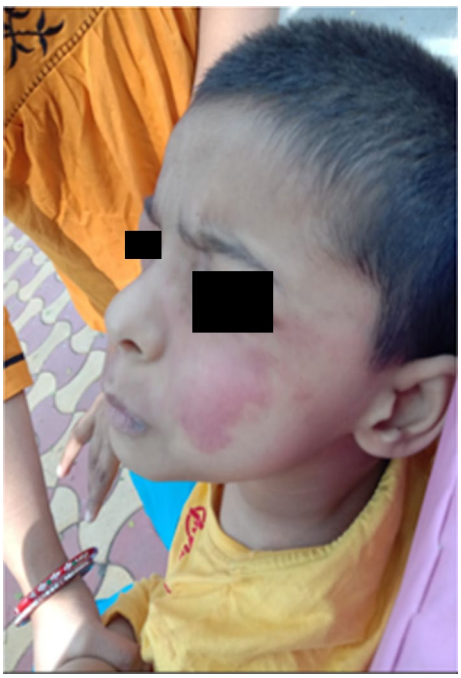

Fig. 2 Showing cutaneous lesions on face

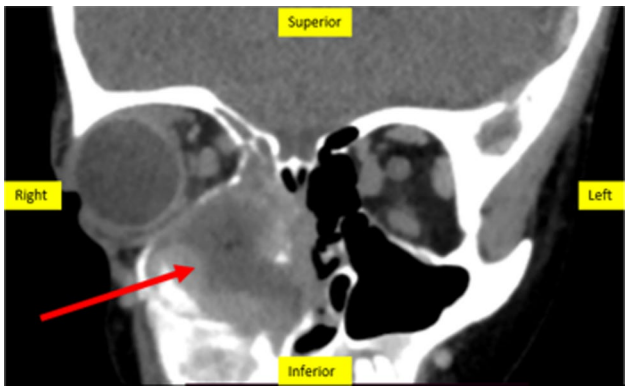

Fig. 4 Coronal view of computed tomography scan showing lesion in right maxilla

and may be pulsatile. Port wine stains are named so due to the deep red hue that they leave on skin or mucosa, and such lesions are characterized by profuse bleeding on trauma. Involvement of the area supplied by ophthalmic division is pathognomic and can result in ocular involvement with glaucoma or blindness [4].

Osteohypertrophy is a benign overgrowth of bone. This osteohypertrophy is described as angiodysplasia, and angiodysplastic syndrome, implies a vascular malformation that is associated with secondary changes including further vascular abnormalities and bone hypertrophy which is frequently observed in Klippel-Trenaunay-Weber (KTW) syndrome involving extremities [5, 6].

According to the distribution of the vascular malformation, manifestations of SWS were divided into the following four parts: (1) cutaneous manifestations, (2) neurological symptoms and signs, (3) ocular manifestations, (4) other manifestations involving oral cavity [7].

Diagnosis and Management

The differential diagnosis includes Rendu-Osler-Weber syndrome, angio-osteodystrophy syndrome, Maffucci's syndrome, Von Hippel-Lindau disease, Trenaunay-Weber syndrome, Bannayan Riley Ruvalcaba syndrome, Divry Van Bogart syndrome and Cobb syndrome.

Diagnosis is based on imaging studies, CSF analysis for elevated protein, skull radiograph for tram line calcification, cranial CT scan for angioma and calcification. MRI is gold standard for diagnosis. Treatment and prognosis depend upon severity of clinical condition. Presence of port wine stain can cause psychological trauma to patient. Port wine stains can be treated by dermabrasion, tattooing, and laser therapy. Cryosurgery can be used to correct lip and other soft tissue deformities. Anticonvulsant drugs can be advised for patients with seizures. Aspirin can be advised for headache and to prevent vascular disease. Eye drops are prescribed for glaucoma [8–10].

The current case presented a clinical dilemma at the time of the boy's presentation because of the uncertainty of diagnosis and aggressive course of disease. On the basis of the location of tumour and territory of cutaneous capillary angioma, one may presume a benign aetiology related to vascular effect (angiodysplasia) rather than neoplastic growth.

Dental management of the patient should be stressed upon with behaviour management and preventive measures. Poor oral hygiene can lead to secondary inflammatory gingival enlargement and high decayed, missing, and filled teeth (DMFT) score. Gingival overgrowth can be managed by proper oral hygiene maintenance and gingivectomy using Nd:Yag laser. Periodontal injection is preferred in these cases to avoid bleeding. Due to risk of haemorrhage, precautions should be taken during surgical procedures. Absorbable hemostatic agents can be placed at extraction socket; endodontic treatment can be performed since angioma may not involve pulpal tissue; over instrumentation should be avoided during periapical instrumentation of root canals; and pulpal bleeding can be controlled by cotton pellet and vasoconstrictors [11].

Although a vascular effect on bone proliferation was originally considered in light of the clinical context of SWS, the rapid local progression of the mass led one to question this diagnosis in favour of a bone tumour. It remains uncertain, however, whether this represents a coincidental osseous neoplasm or an associated fibro-osseous neoplastic transformation related to cutaneous vascular malformation [12].

Treatment is aimed at correcting or preventing functional problems and achieving normal facial aesthetics. Conservative shaving or osseous contouring is recommended; if at all possible, this treatment should be postponed until after lesion growth subsides. If continued growth is observed after conservative treatment, periodic contouring may be performed until a static phase is reached. This repeated contouring operation might produce less total morbidity than a single massive resection. Conversely, orbital hypertelorism, dystopia, exophthalmos, or grotesque orbitofacial deformity cannot be corrected by conservative "contouring"; they can only be corrected by radical excision and reconstruction. Radiation therapy of fibrous dysplasia is mentioned only to be condemned because of its questionable therapeutic value and possible relation to subsequent malignant transformation [13–16].

Symptomless fibrous dysplasia may be found accidentally in radiographs or computed tomography scans. Surgery on these lesions is not considered unless there is evidence of progression during annual follow-up or if symptoms indicating a mass appear [17].

Conclusion

A patient of SWS may present with varying symptoms and rarely an association of OF may be a possibility as reported here. Clinical evaluation of such patients must be extensive prior to surgical workup. Surgical excision of lesion (OF) in patients with Sturge–Weber syndrome is challenging due to high risk of haemorrhage. Precautionary measures must be taken to control haemorrhage and complications during surgical procedures. Most importantly, each patient may present with variable symptoms and clinical findings, thus keeping this in mind each patient would require long term management and care, the same must be customized to their needs and sites of involvement.

Declarations

Conflict of interest The authors declare that they have no conflict of interest.

Research Involving Human Participants and/or Animals The clinical case report involves human participant (s).

Informed Consent Informed consent was obtained from the individuals participating in the study.

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