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Fibrous Dysplasia of Maxilla with Port Wine Stain: A Case Report

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Case Report

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Abstract: Fibrous dysplasia (FD) is an uncommon skeletal disorder in which normal bone is replaced by abnormal fibro osseous tissue. Mainly, Fibrous Dysplasia is found in children, and by adulthood it usually becomes quiescent. Our case showed Fibrous dysplasia of 9-year-old female child with port wine stain on the right side of mid face since birth. Our evaluation was that growth in childhood had achieved the progressive stage. We presented all radiographic and clinical images of this case that emphasized the diagnosis of the disease.

Keywords: Fibrous Dysplasia, Maxilla, Monostotic, Polyostotic, Recontouring.

INTRODUCTION

Fibrous dysplasia (FD) is an idiopathic skeletal disorder in which the trabecular bone is replaced and distorted by poorly organized, structurally unsound fibro-osseous tissue. The lesion is classified into two forms: Monostotic (75-80%) and polyostotic. A distinct form of Polyostotic FD, known as McCune-Albright Syndrome, is accompanied by cutaneous pigmentation and sexual precocity, and this occurs almost exclusively in women. Typical radiographic appearance shows an expanded osseous lesion having poorly defined margins covered by a thin "eggshell" cortex and lacking periosteal new bone formation. Here, we are presenting two case reports of FD involving the maxilla. Fibrous dysplasia (FD) is a non-malignant condition caused by post-zygotic, activating mutations of the GNAS gene that result in inhibition of the differentiation and proliferation of bone-forming stromal cells and leads to the replacement of normal bone and marrow by fibrous tissue and woven bone [1].

The disease may present in a monostotic or polyostotic form, affecting one or multiple bones, respectively. Mainly, FD is found in children, and it usually becomes dormant by adulthood [2]. The age range of patients with FD has been reported to be between 20 and 70, with the majority diagnosed in their 30s [3]. Despite recent advances in the understanding of the natural history and molecular abnormalities of FD, many questions remain regarding its progression and management [4]. Although most cases of FD are self-limiting [5] and hamartomatous, some cases of FD do not go into dormancy at the end of adolescence [6] and may be activated or reactivated in adulthood in response to a life event, such as pregnancy [7, 8]. Because some cases of FD in elderly people have also been reported [9-11], we directed attention to the question of whether a long-term case could show continuous progress or would cease in the quiescent stage. We also examined the incidence of FD transformation into malignancy. Our case showed Fibrous dysplasia of 9-year-old female child with port wine stain on the right side of mid face since birth. Our evaluation was that growth in childhood had achieved the progressive stage. We presented all radiographic and clinical images of this case that emphasized the diagnosis of the disease.

CASE REPORT

A 9-year-old female patient reported with her parents with the chief complaint of painless swelling and discoloration on the right side of face with excessive upper teeth and gum show on the right side since childhood. the disease progressed slowly as time advanced which started with a pea size and gradually increased to attain the present size. There was no history of trauma, paresthesia, and difficulty in chewing food, and it was not associated with any other symptoms. Extra orally (Figure-1), a diffuse swelling (4 × 5 cm approximately) was seen, superioinferiorly starting from 2 cm below the infraorbital margin to the line joining the commissure and the ear lobe, and mediolaterally starting from the left side of nasal

septum, causing obliteration of left nasolabial fold, to maxilla on the right side, which was bony hard and nontender in nature. Intraorally, the swelling started form the central Incisor region on the left side crossing the midline extending from 21 to 18 tooth region with palatal extensions and ectopically erupted tooth in the involved region, with expansion of the buccal cortical plate and obliteration of vestibule. Overlying mucosa was smooth and it was bony hard and nontender (Figure-2). Presence of port wine stain on the right region of face (Figure-3). Provisional diagnosis of fibro-osseous lesion of right maxillary region was made. Teeth in the vicinity of the lesion were vital. Complete hemogram showed all the parameters were within normal limits. Serological investigations including serum calcium, serum phosphorus, and alkaline phosphatase (ALP) were also within normal range. Orthopantomogram (OPG) (Figure-4) and PNS X ray (Figure-5) revealed and a homogeneous ground-glass radio-opaque pattern in the periapical areas involving the maxillary sinus and excessive radiopacity and increase in size of maxilla. Higher radiographic investigations (CT Scan) with axial coronal and sagittal sections were done to rule out the extensions (Figure-6) which revealed homogeneous, granular radiopacity of right maxillary alveolar bone with expansion of buccal cortical plate extending from 21 to 18 region, giving a granular or ground-glass appearance. On the palatal side, homogeneous radio-opacity partially covered the greater palatine foramen. Margins of the lesion blended with adjacent areas. Angiogram was taken of the involved area to rule out any vascular malformations which was negative (Figure-7) Chest X ray was taken as patient was planned to be operated under General anesthesia which was clear (Figure-8). Surgical recontouring of maxilla was done and histopathological examination revealed a connective stroma which was cellular and fibrous with plump cells. It was intermixed with bone of varying sizes and shapes confirming Fibrous dysplasia. The histologic features were suggestive of a fibro-osseous lesion. After the clinical features and radiological features were correlated, it was diagnosed as Fibrous dysplasia. Patient was discharged after 3 days and recalled after 2 weeks for follow up. On follow up she was happy with her appearance in the post-operative follow up after 2 weeks (Figure-9).

Fig-1: Clinical Picture of Patient

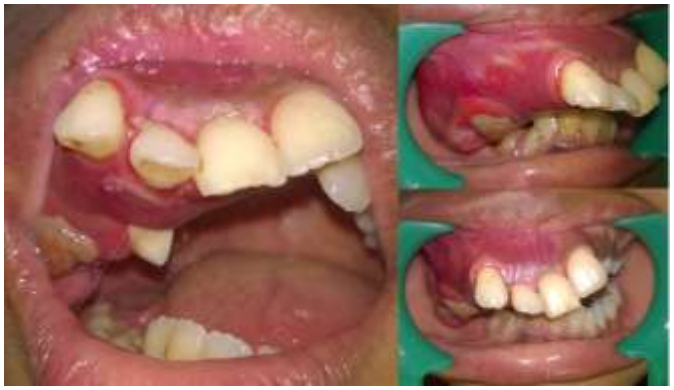

Fig-2: Intra-oral Pictures

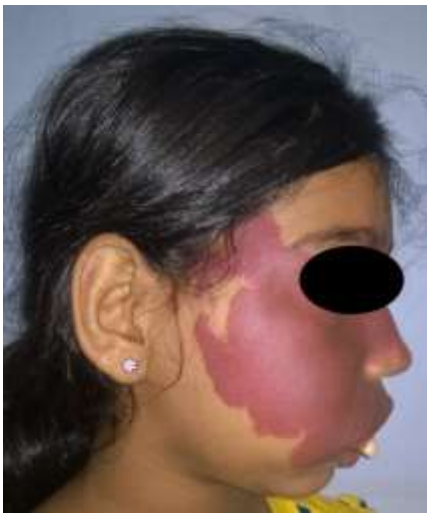

Fig-3: Picture showing Port wine stain on right side of face

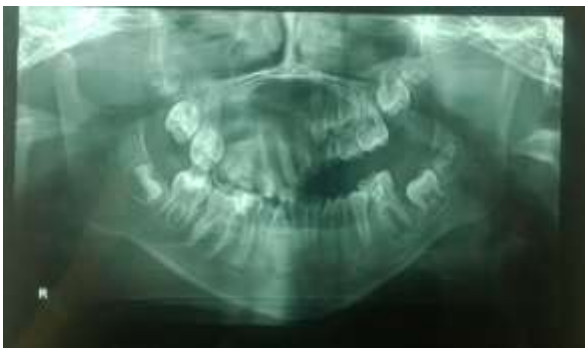

Fig-4: OPG X ray

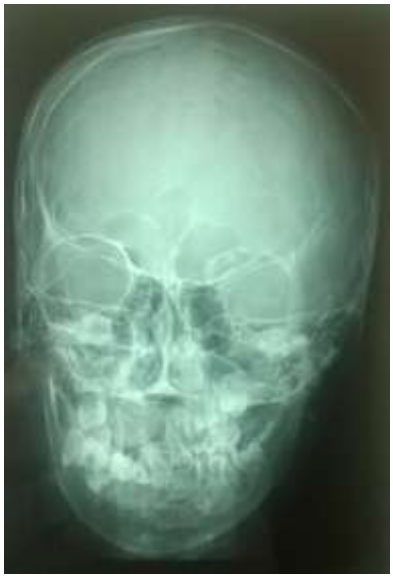

Fig-5: PNS X ray

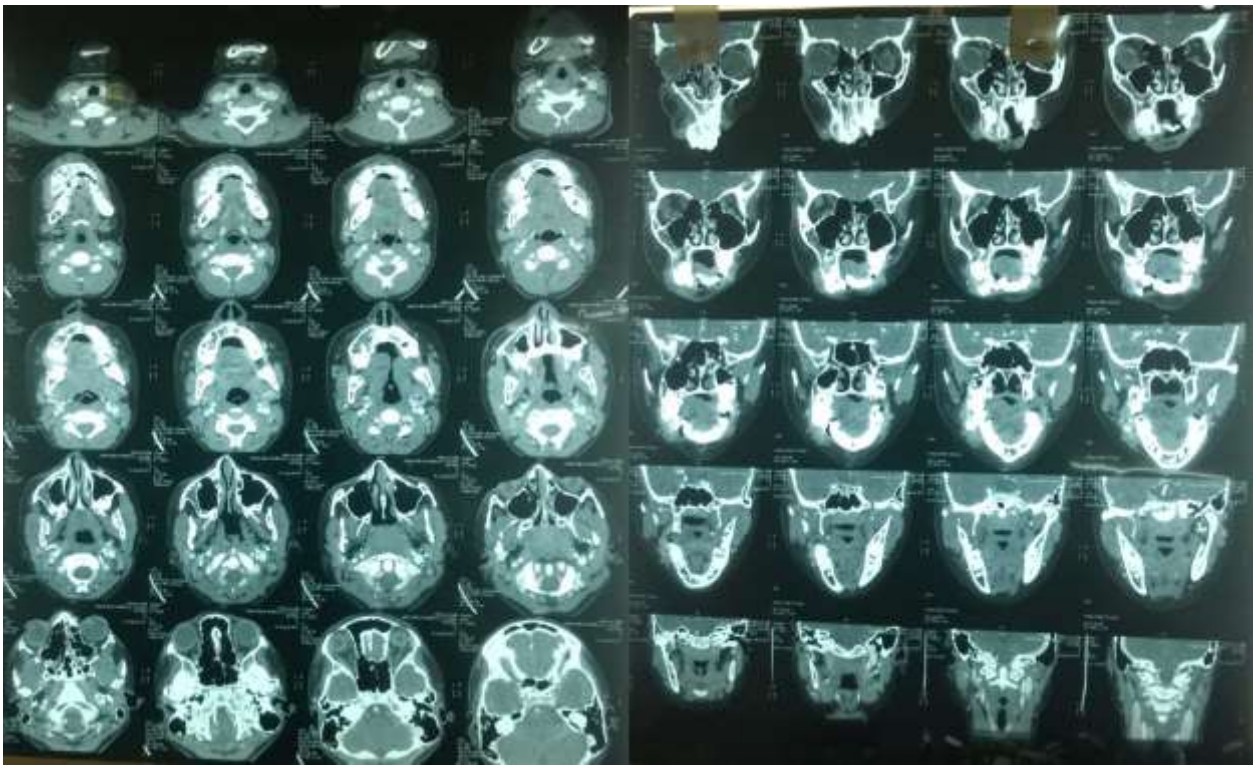

Fig-6: CT Scan

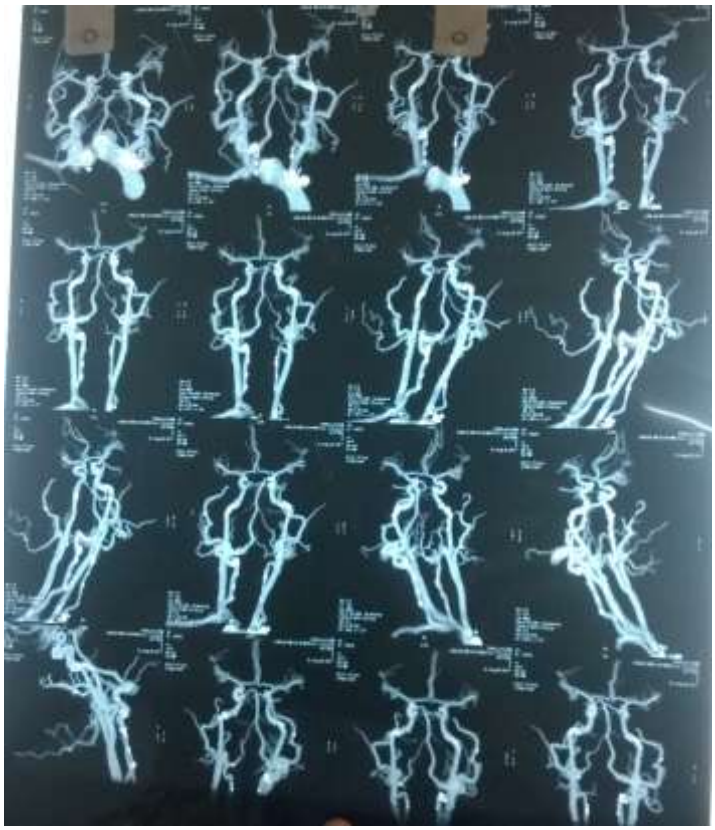

Fig-7: Angiogram

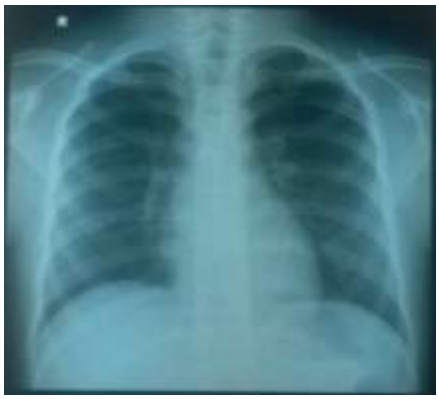

Fig-8: Chest X ray

Fig-9: Post-operative follow up clinical picture after 2 weeks

CONCLUSION

Fibrous dysplasia (FD) is a disturbance of bone metabolism that is classified as a benign fibro-osseous lesion. The fibrous connective tissue containing abnormal bone replaces normal bone. FD is a benign intramedullary fibro-osseous lesion. It is a sporadic benign skeletal disorder that can affect one or multiple bones, and the latter may form part of syndromes. FD is also attributed to gene mutation. Knowing all these features of FD it is very important to diagnose and treat such case promptly to increase the quality of life of patients.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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CONFLICTS OF INTEREST

There are no conflicts of interest.

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