

## Fibrous dysplasia of the clivus, a case report

Kamour A<sup>1</sup>, Lewis D<sup>2</sup> and Ismail A H<sup>1</sup>

(1) Department of elderly medicine and (2) Department of Radiology, Royal Albert Edward Infirmary, UK

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### Introduction

Fibrous dysplasia (FD) of the clivus, first described by Lichtenstein in 1938, is a very rare developmental, non-hereditary disorder caused by abnormal proliferation and maturation of fibroblasts resulting in replacement of mature bone by weak and immature bone.

### Case report

A 26-year-old otherwise fit and healthy Caucasian female presented to the Emergency Department with headache for 3 days, recurrent and getting worse. Neurological examination was normal. Blood investigations were essentially normal. Head CT scan showed slightly expanded clivus with ground glass density ( fig 1 ). MRI revealed slight expansion with focal signal alteration within the clivus, being hypointense on T1W images and FLAIR images and showing slightly increased signal intensity on T2W images (figure 2).

While in hospital the patient's symptoms resolved with conventional painkiller treatment. Based on the current evidence available we opted for a conservative treatment and regular follow-ups and up to the day of writing this report, about eight months later, the patient is still on the same management plan.

### Discussion

In 70% of cases FD affects a single bone, Monostotic FD, with the frontal, sphenoid, ethmoid, orbit, zygoma, maxilla, mandible, and temporal bones being the common sites affected in craniofacial bones. However, monostotic fibrous dysplasia of the clivus is extremely rare [1, 2]. In 30% of cases FD is polyostotic affecting several bones, where the craniofacial bones are mainly affected.

McCune-Albright syndrome constitutes a Triad of polyostotic FD, skin hyperpigmentation, and endocrine dysfunction (hyperthyroid, precocious puberty, Cushing syndrome, acromegaly etc)

FD of the Clivus can be either asymptomatic or present with headache but in severe cases cranial nerve involvement may occur. On X-ray, three patterns of FD are identified: Cyst-like, Sclerotic and Mixed "Pagetoid" [2].

CT SCAN shows the characteristic appearance of thinning of the cortical bone, expansion of the affected area with "ground glass" density. [2].

MRI appearance varies between Low to intermediate T1 and low to high T2 signal.

The MRI appearance may not be as specific as the CT. however, the key findings In favour of FD on MRI are:

1- expansion of the involved bone with intact cortical outline, along with a varied degree of contrast enhancement within the lesion and not along the adjacent thickened dura.

2- FD in its early stages shows T2 hyperintensity but once the lesion becomes mineralized, it will appear hypointense on T1W and more so on T2W MR Image [3,5].

The variation in MRI appearances may also be due to the overall cellularity, collagen content, extent of bone trabeculae, and cyst formation. [3, 5, 7].

It has been suggested that it is possible to obtain a correct diagnosis of fibrous dysplasia without the need for a biopsy. [3, 6]. However, biopsy might be needed if the diagnosis is doubtful or malignant transformation is suspected [4].

The management of fibrous dysplasia is not surgical unless it causes unacceptable or progressive deformity, cranial nerve compromise; severe headache, or development of a malignancy [1, 6]. Usually small, non-expansile solitary lesions will remain unchanged. Although the prognosis of fibrous dysplasia is generally good, malignant degeneration and aggressive behaviour have been described. The incidence of malignant transformation is highest for monostotic craniofacial lesions (0.05%). Osteosarcoma, fibrosarcoma, and chondrosarcoma are the malignancies reported in the literature.

A CT follow-up would be more appropriate for detection of any subtle changes in the morphology and internal structure of the lesion [6].

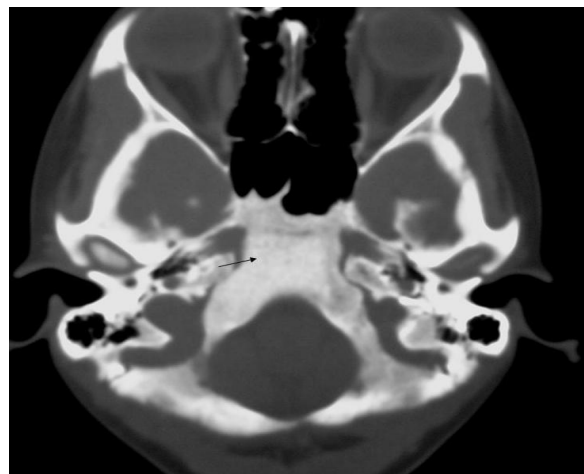
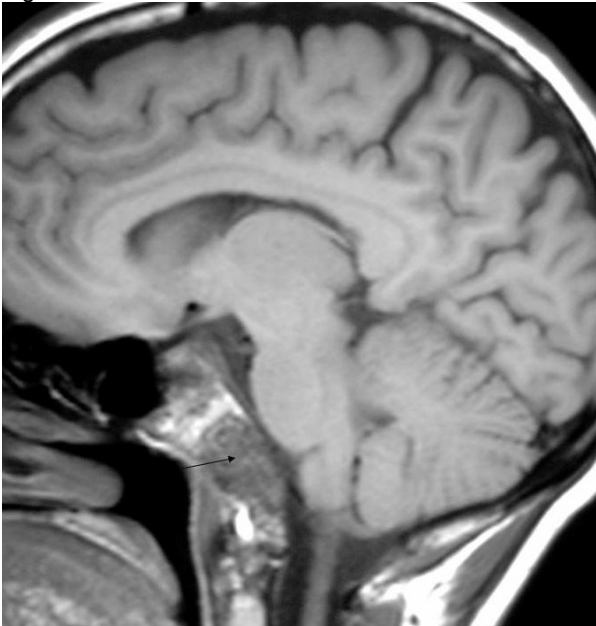
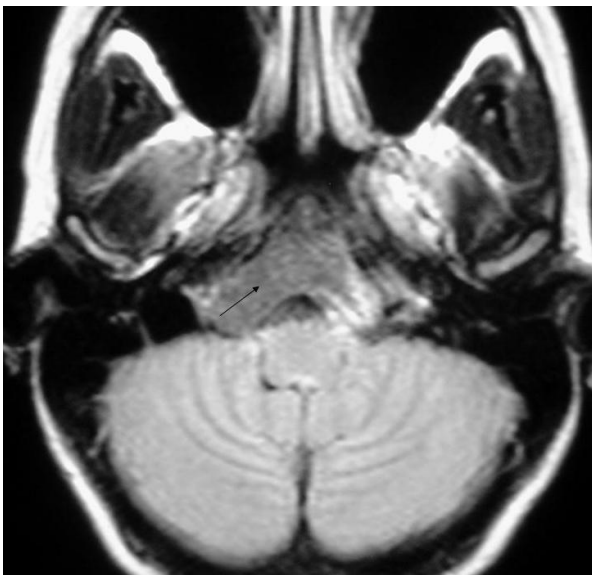


Figure 1: CT brain, bone window, shows expanded clivus with ground glass density.

Figure 2: MRI brain



A- Sagittal T1W image shows low signal expanding clivus lesion.



B- Axial FLAIR image shows low signal well defined lesion in the clivus.

### Conclusion

Fibrous dysplasia of the clivus is a rare disease, may be asymptomatic or may present with vary neurological manifestations. The diagnosis depends mainly on the typical CT-scan picture and biopsy is not needed unless malignancy is suspected or diagnosis is doubtful. Management in most cases is conservative unless unacceptable symptoms or complications have developed

### References

1. Levy ML, Chen TC, Weiss MH .Monostotic fibrous dysplasia of the clivus. J Neurosurg. 1991; 75:800-803
- 2.Sato K, Kubota T, Kaneko M, Kawano H, Kobayashi H .Fibrous dysplasia of the clivus. Surg Neurol1993; 40:522-525
3. M. Sirvanci, K. Karaman, L.Onat.Monostotic fibrous dysplasia of the clivus: MRI and CT findings. Diagnostic NeuroradiologyDOI 2002; 10.1007/s00234-002-0833-8
4. D.Wayne Ham, K.Pitman.Fibrous dysplasia of the clivus and sphenoid sinus: case report: military medicine 1998; 163,3:186
5. Utz JA, Krandsdorft MJ, Jelinek JS, Moser PR, berry BH. MR appearance of FD .j comput assists tomogr. 1989; 13:845-851
6. Badih A, Ossama AL. fibrous dysplasia of the clivus.neurosurgery.2003; 52:318-323
7. Norris MA, Kaplan PA, Pathria M, Greenway G.Fibrous dysplasia: magnetic resonance imaging appearance at 1.5 tesla. Clin Imag 1990; 14:211-214