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## Case Report

# Fibrous dysplasia of the clivus – A case study and literature review <sup>☆</sup>

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## ABSTRACT

Fibrous dysplasia is a benign, congenital skeletal disorder which leads to the formation of fibro-osseous intramedullary bone lesions. Clival fibrous dysplasia is a rare variant which commonly presents asymptotically with no findings on examination and is often picked up incidentally on radiological investigation. A 39-year-old female presented with a sudden onset headache of 3 days' duration alongside diplopia and right lower limb weakness upon examination. Computerized tomography head scan revealed an expansile clivus with a ground-glass appearance, magnetic resonance imaging brain scan revealed a predominantly hypointense signal on T1- and T2-weighted images and subsequent whole-body bone imaging confirmed the diagnosis of monostotic clival fibrous dysplasia. This case highlights the importance of considering monostotic clival fibrous dysplasia as a differential diagnosis in patients presenting with sudden onset symptoms of headache alongside cranial and peripheral nerve involvement, when other more sinister causes have been excluded.

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## Introduction

Fibrous dysplasia is a rare congenital skeletal disorder which leads to the formation of benign fibro-osseous intramedullary bone lesions throughout the skeletal system [1,2]. The majority of cases are monostotic, that is, involving single bones, accounting for between 70% and 85% of cases. Several bones

can also be involved, termed polyostotic [3]. The lesions are commonly found in the craniofacial bones, long bones, pelvis and ribs and reportedly account for between 5% and 7% of all benign bone tumors. Fibrous dysplasia can present at any age with the bulk of cases being identified by the age of 30 with no known gender bias [2]. Monostotic fibrous dysplasia typically presents between 20 and 30 years of age, commonly in the femur or ribs and is often asymptomatic and inci-

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dentally picked up on radiological imaging for an unrelated indication [2–4].

Postzygotic activating mutations in *GNAS* are the driving force behind the pathophysiology of fibrous dysplasia. *GNAS* encodes the  $\alpha$ -subunit of the  $G_s$  stimulatory protein and 2 potential mutations can occur which lead to interruption of the intrinsic GTPase activity of  $G_s\alpha$ . This leads to receptor activation and consequent inappropriate cyclic AMP-mediated signaling. [5,6] This mutation occurs early on in embryogenesis before the formation of the endoderm, mesoderm and ectoderm. Both bone resorption and formation are affected due to the consequent effect of the relevant gene mutation on skeletal stem cells. Activation of  $G_s\alpha$  leads to the formation of bone marrow stromal cells which cannot differentiate into normal osteoblasts, adipocytes and hematopoiesis-supporting cells. Consequently, the normal cancellous bone and its marrow are replaced by proliferating bone marrow stromal cells, which form structurally weak discrete fibro-osseous lesions [6,7]. These lesions have a bone matrix deficient in osteopontin and bone sialoprotein which is a marker of osteoblastic cell differentiation and necessary for mineralization. The consequent lack of mineralization is referred to as localized osteomalacia [8].

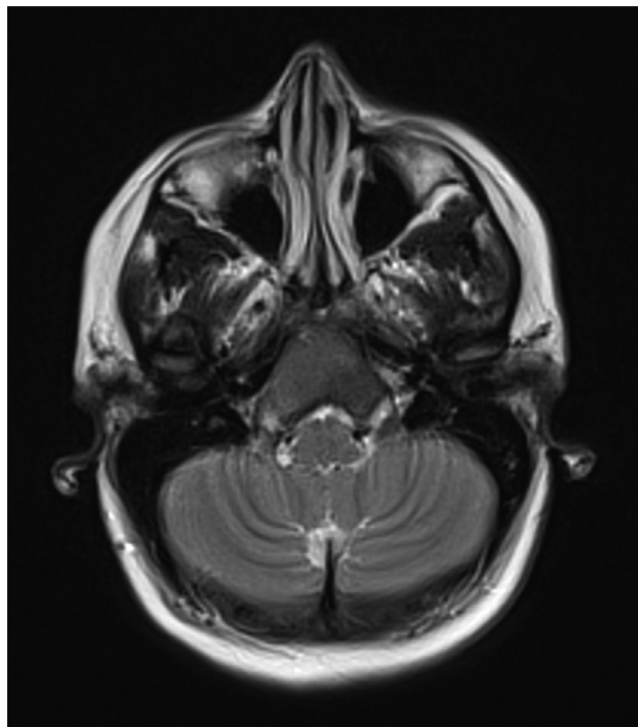
We present a case of fibrous dysplasia of the clivus in a 39-year-old female and go on to discuss symptomatology of clival fibrous dysplasia as well as radiological findings. We compare our case with other previously reported cases of fibrous dysplasia of the clivus.

## Case report

A 39-year-old female presented to the emergency department with a 3-day history of severe headache. This began during sexual intercourse at 2 AM, reported as 9/10 in severity and was maximal at onset, located all over the top of the scalp and in the occipital region. The symptoms eased after 1 hour with headache intensity dropping to a reported 5/10 and remaining persistent since that time.

Upon examination there was isolated right-sided lower limb weakness, which intermittently worsened with straining and coughing, along with some associated diplopia. The patient underwent an initial computerized tomography (CT) head scan without contrast to rule out subarachnoid hemorrhage. This was originally reported as normal, showing no signs of any acute intracranial event, no space-occupying lesions, and normal appearances of brainstem and cerebellum. CT angiogram scan was also done to look for any signs of subarachnoid hemorrhage and was reported as normal.

Subsequently, the patient then had a magnetic resonance imaging (MRI) head scan which showed an incidental finding of abnormal enlargement and signal change within the clivus. On T1-weighted sequences there was a predominantly low signal with a few areas of intermediate signal noted. A similar pattern was seen on T2-weighted images, predominantly with areas of low signal attenuation (Figs. 1 and 2). Upon retrospective review of the original CT head scan, and looking at the cervicomedullary junction and bone windows, crowding at the foramen magnum with decreased cerebrospinal fluid



**Fig. 1 – Axial T2-weighted MRI sequence showing predominantly low signal, with few areas of intermediate signal, in the expansile clival lesion. A similar pattern was seen on T1-weighted images with predominantly areas of low signal attenuation within the lesion.**

spaces were identified together with an expansile appearing clivus, which exhibited a ground-glass appearance with loss of trabecular pattern (Figs. 3 and 4). This appeared to be contributing towards narrowing of the foramen magnum.

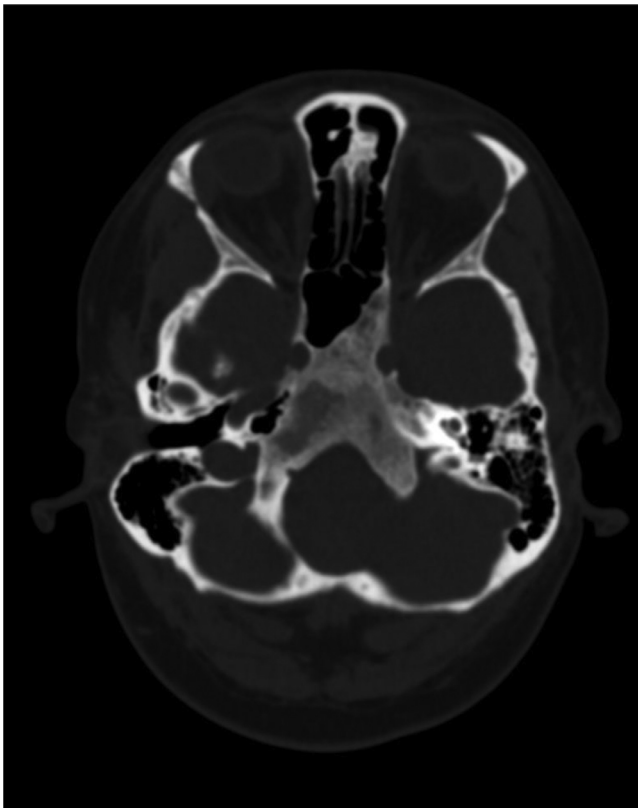
The differentials at this point were fibrous dysplasia, chordoma and plasmacytoma. Chordomas are often more destructive and aggressive-looking tumors whilst fibrous dysplasia lesions are smoother and respect bony outlines, which was seen in this case. Given the ground-glass appearance, plasmacytoma was unlikely as these are isodense to brain parenchyma and often present in an older age group. To differentiate between monostotic or polyostotic fibrous dysplasia, a bone scan in the first instance along with neurosurgical opinion was advised.

Whole body bone imaging with technetium 99m-methyl diphosphonate showed a solitary focus of increased uptake in the midline skull base, corresponding to the expansile lesion seen on the MRI and CT scans, which had demonstrated a ground-glass appearance with no bony destruction on CT scan. No other sites of abnormal tracer uptake were seen (Fig. 5). Consequently, we were able to rule out other differentials, such as malignancy, and come to the conclusion that this is monostotic fibrous dysplasia of the clivus based on clinical and radiological features.

In terms of management, the patient will undergo follow up scans over the course of the next few years to ensure no progression or malignant transformation of the clival lesion.



**Fig. 2 – Sagittal T2-weighted MRI sequence showing predominantly low signal, with few areas of intermediate signal, in the expansile clival lesion.**



**Fig. 3 – CT head scan axial view showing ground-glass appearance of the well-defined, expansile clival lesion.**

## Discussion

Fibrous dysplasia of the clivus is a rare disease with 43 reported cases in the existing literature [7,9–32]. The clivus is a base of skull bone formed from the basisphenoid and basiocciput. As we progress into adulthood, clival marrow changes from low/intermediate T1 signal to a high T1 signal. The presence of a low T1 signal in adults is an indicator of disease with common clival pathology being chordoma, myeloma, metastases, chondrosarcoma, nasopharyngeal carcinoma, pituitary macroadenoma, cholesteatoma, mucocoele and of course fibrous dysplasia [33].

When looking at the 43 previously reported cases, the mean age of diagnosis was 35-years-old for male patients and 43-years-old for female patients, which aligns with previously reported figures [7,9–32]. Our own patient, who was diagnosed at age 39-years-old, also fits into this bracket.

As mentioned previously, the majority of cases of fibrous dysplasia are monostotic, and our review of previous cases supports this for clival lesions also [3,7,9–32]. Of the 43 reported cases of clival fibrous dysplasia, only 7 underwent radionuclide scanning with 5 (71%) of these patients having monostotic and 2 (5%) having polyostotic disease. Thirty-six (84%) of the patients did not have any reported radionuclide scanning and of these, 20 (56%) had monostotic and 5 (14%) had polyostotic disease of the clivus based upon their CT and/or MRI findings alone, with 11 (31%) patients scan results not being mentioned. Monostotic was defined as purely clival involvement whilst polyostotic referred to additional bone involvement. Consequently, based upon CT, MRI, and radionuclide results in conjunction there were 25 (58%) monostotic

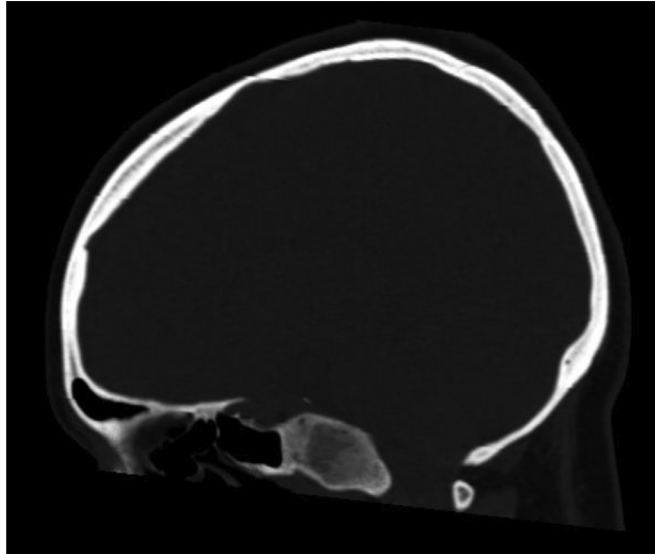


Fig. 4 - CT head scan sagittal view showing ground-glass appearance of the well-defined, expansile clival lesion.

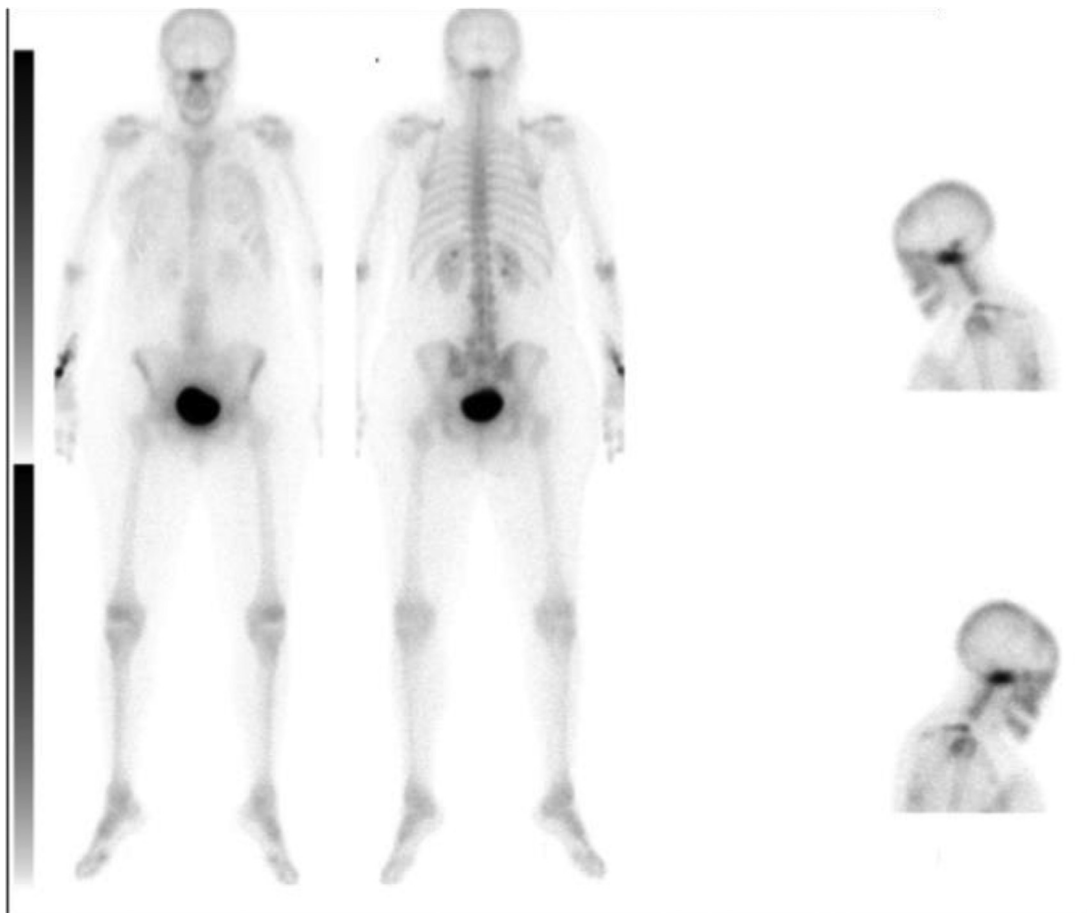


Fig. 5 - Whole body bone imaging showing involvement confined to clivus. There is increased uptake in the clival lesion with no evidence of multifocal disease.

cases and 7 (16%) polyostotic cases of fibrous dysplasia of the clivus [7,9–32]. Our patient underwent a full workup of scans including CT (Figs. 3 and 4) and MRI (Figs. 1 and 2) scans of the brain as well as a nuclear medicine bone scan (Fig. 5) and was found to have monostotic clival fibrous dysplasia.

Symptoms of clival fibrous dysplasia can vary greatly and often patients can be completely asymptomatic. Signs and symptoms of fibrous dysplasia usually come about due to progressive bone deformation over many years and can differ depending upon the site and extension of the lesion. In craniofacial fibrous dysplasia patients often report a unilateral, painless expansion of the skull which can result in intracranial extension and consequently central neurological manifestations, neurovascular entrapment, orbito-sino-nasal obstruction, proptosis and cosmetic deformity to name but a few. In base of skull fibrous dysplasia patients can often present with cranial nerve symptoms often affecting facial, auditory, trigeminal, olfactory and visual function due to thickening and narrowing of neural canals and foramina [7]. Symptoms were reported in 34 of the 43 previous cases of clival fibrous dysplasia. Of these 34 (79%), the most common symptom was headache, with 22 (65%) patients reporting this as a symptom [7,9–32]. A localized occipital headache has previously been associated with extension of disease to the occipital condyle. [7] Cranial nerve symptoms were seen in 8 (24%) patients with 5 (15%) patients being asymptomatic. Timeframe of symptom onset to diagnosis was only mentioned in 11 of the previously published cases and our review shows that of these 11 most patients usually present after having had symptoms for several months, with 8 (73%) fitting into this category and only 2 (18%) having symptoms for years [7,9–32]. Our patient's presentation with a headache fits with the classical symptomatology of clival fibrous dysplasia, however, the timeframe of her symptoms makes this a noteworthy case. Our patient only had symptoms lasting for only 3 days and from our review there was only 1 (9%) other patient, reported by Kamour et al., who presented in this manner, also with headache lasting for 3 days [26]. Our patient's presentation of clival fibrous dysplasia is therefore extremely rare in terms of how suddenly she developed her symptoms.

Examination of patients with clival fibrous dysplasia is often unfruitful with our review showing that, of the 28 (65%) patients who had examination findings reported, 22 (79%) had no signs elicited on examination. Cranial nerve deficit was seen in 3 (11%) patients whilst nasal signs and masses were noted in 2 (7%) patients each [7,9–32]. There was only 1 (4%) reported case, by Iseri et al, of peripheral nerve signs where their patient had wasting and weakness of upper limbs, pyramidal signs in the lower limbs, and dissociated sensory loss C4–C8 dermatomes accompanied by a mass and cranial nerve signs, although this was a case of polyostotic fibrous dysplasia of the skull base with an aneurysmal bone cyst of the occipital bone [21]. These findings are similar to our patient who also had peripheral nerve signs and cranial nerve deficit on examination with right-sided lower limb weakness along with intermittent diplopia. From our review of the previous cases this makes our case only the second reported case of clival fibrous dysplasia presenting with cranial nerve deficit and limb weakness [7,9–32]. Our case is unique in that our patient only had unilateral limb weakness, as well as the unusual phenomenon

of this worsening on straining and coughing, which has not been previously reported in a case of clival fibrous dysplasia.

Our patient underwent an extensive workup of scans and no other identifiable cause for her symptoms and examination findings were found. Consequently they were put down to monostotic fibrous dysplasia of the clivus. As previously mentioned, cranial nerve symptoms as a result of clival fibrous dysplasia are commonly associated with thickening and narrowing of neural canals and foramina, and mass effect from the lesion itself can also play a part [7,21]. In the case discussed by Iseri et al they report that involvement of the jugular foramen and fossa, hindbrain herniation and syringobulbia were likely to be the cause of the patient's array of cranial and peripheral nerve symptoms [21]. Our patient's findings were very different from this, with only crowding and narrowing at the foramen magnum, decreased cerebrospinal fluid spaces and an expansile clivus being identified. It is possible that these findings, together with mass effect from the lesion itself, were behind the pathoanatomical mechanism behind the patient's symptoms and examination findings. However, due to the relative paucity of similar case presentations, it is difficult to be certain as to the exact mechanism behind the patient's presentation and further research would be necessary to adequately ascertain this.

MRI scanning is often the imaging modality of choice for base of skull lesions and fibrous dysplasia is often picked up incidentally and regularly misinterpreted as a tumor. Classically, T1-weighted images in fibrous dysplasia display low-intermediate signal intensity which is dependent on the ratio of fibrous tissue to mineralized matrix, with those lesions with highly mineralized matrix showing lower signal intensity whilst those with high fibrous tissue content display intermediate signal intensity. T2-weighted signal intensities are often more variable with some lesions with high amounts of fibrous tissue showing high signal intensity which is believed to be due to this fibrous tissue being metabolically active. This explains why fibrous dysplasia lesions usually have intense enhancement after injection of contrast, as they are highly vascularized [34]. Of the previously reported 43 cases, T1-weighted images were mentioned in a total of 30 (70%) cases and of these, 29 (97%) showed the classical, purely hypointense signal. Twenty-nine (67%) of the cases mentioned T2-weighted images with 19 (66%) showing only a hypointense signal. MRI contrast enhancement was seen in 100% of the 21 (49%) cases which reported administration of IV contrast [7,9–32]. Our patient's MRI scan displayed the classical features of fibrous dysplasia with a predominantly hypointense signal on T1-weighted images, with a few scattered areas of intermediate signal, and T2-weighted images were principally hypointense (Figs. 1 and 2). There was an absence of any high signal intensity regions, indicating a lesion of low metabolic activity.

Diagnostic uncertainty of fibrous dysplasia on MRI arises when the classical finding of low signal intensities on T1- and T2-weighted images are not seen, yet MRI contrast enhancement is still evident. This finding is often labelled as a neoplasm. In this scenario it is advised that CT scanning should be performed and it has been previously reported that 56% of CT scans show ground-glass opacity, although other variants do exist such as homogeneously dense and cystic [34].



Expansion of the affected bone and a thin cortex are also classical features of fibrous dysplasia [7]. Of the 43 cases of clival fibrous dysplasia, 29 (67%) of them reported their CT findings and 25 (86%) of these mentioned a ground-glass appearance when describing the changes seen. [7,9–32] Our patient's scans demonstrated the classical ground-glass appearance on CT with an expansile appearing clivus (Figs. 3 and 4). This helped confirm the diagnosis despite absence of contrast enhancement on CT angiogram, which was unusual. Furthermore, this case emphasizes the importance of looking at the cervicomedullary junction and bone windows to evaluate the clivus as this was initially not reported and only detected on retrospective review.

The differential diagnoses that were postulated at the time of the initial CT included chordoma and plasmacytoma, 2 diseases that arise from within the clivus. Chordomas have insidious onset and present with headache meaning they can be confused with fibrous dysplasia of the clivus initially. Characteristically chordomas have high signal intensity on T2-weighted images, often with heterogeneous contrast enhancement, allowing them to be differentiated. Plasmacytomas are usually moderately hyperintense on T2-weighted images and typically have an invasive outline on CT. Looking at other potential differentials, chondrosarcomas usually occur at the petroclival synchondrosis hence are sited in a paramedian location and often present with cranial nerve palsies, which overlaps with fibrous dysplasia symptomatology. They can be differentiated using MRI where T2-weighted images will be of high signal. The chance of a mass in the clivus being metastatic is low without a known primary, but in those with previously identified malignancy the probability increases significantly. When thinking of pathology arising from adjacent structures: pituitary macroadenomas usually have their epicentre in the sella turcica; nasopharyngeal cancers originate from the nasopharyngeal mucosal space and often present with eustachian tube dysfunction causing otitis media; cholesteatoma symptoms occasionally crossover with those of clival fibrous dysplasia with facial nerve palsy, tinnitus, vertigo and headache but can be differentiated on MRI as they are usually hyperintense on T2-weighted images and nonenhancing; mucoceles classically present with cranial nerve and headache symptoms with variable densities on CT and variable signal intensity on MRI but characteristically have a filled and expanded sinus on imaging [33].

Clival fibrous dysplasia is usually managed with clinical observation as the majority of patients are asymptomatic [7]. Although small, nonexpansile solitary lesions commonly remain unchanged, there have been reports of malignant degeneration and aggressive behavior with the rate of malignant transformation of monostotic craniofacial lesions being 0.05%. Consequently, follow-up usually entails yearly CT scanning of the cranial base along with clinical evaluation. Biopsy is often not required if the CT and MRI images are typical for fibrous dysplasia [29]. Surgery is usually reserved for cases where there is progressive or unacceptable deformity, cranial nerve involvement, extension to the condyles, severe pain or malignant transformation [7,29]. In cases with solitary clival lesions without condylar extension, the lesion can be removed with a conventional or endoscopic approach, whilst occasional lesions will require more extensive procedures [7].

## Conclusion

We present a rare case of fibrous dysplasia affecting the clivus and review the literature of previously reported cases. The diagnosis is often established upon uncovering the classical radiological findings on CT and MRI scans as well as the presence of classical symptomatology and examination findings. Our case emphasizes the need to properly evaluate the cervicomedullary junction with appropriate windowing and consider clival fibrous dysplasia as a differential diagnosis for patients presenting with headache and/or cranial nerve symptoms, when other more common causes have been excluded. We also highlight that although symptomatic patients classically present with longstanding symptoms, it is important to appreciate that patients can present with sudden onset symptoms, masquerading as more sinister diagnoses. It is crucial to differentiate fibrous dysplasia of the clivus from other aggressive clival lesions as management can differ significantly. Clival fibrous dysplasia is generally managed conservatively, with follow up imaging to ensure there is no malignant transformation.

## Patient Consent Statement

Written, informed consent was obtained from the patient for the publication of their case.

## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2020.11.019](https://doi.org/10.1016/j.radcr.2020.11.019).

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