

# SPHENOID BONE DYSPLASIA -A CASE OF UNRESOLVED HEADACHE IN PRIMARY CARE

Abdul Hadi Said, Tg Putri Syarizan\*

*Kulliyah of Medicine, International Islamic University of Malaysia*

\*Corresponding Author's Email: [tengkuputrisyarizan@gmail.com](mailto:tengkuputrisyarizan@gmail.com)

## ABSTRACT

Headache is a very common presenting symptom in patients seeking treatment in primary care. Although most of it is primary headache, physicians may easily miss the diagnosis of secondary headache. Therefore, we need to equip ourselves with adequate knowledge together with a good and thorough physical examination to avoid missing a diagnosis of secondary headache. In addition to that, for a patient presenting with chronic headache, it is very important for us to always review our diagnosis and our management. We reported a case of sphenoid bone fibrous dysplasia who presented with severe recurrent headache despite being given treatment, analgesics and even propranolol for migraine. Fibrous dysplasia accounts for approximately 5 to 7 percent of benign bone tumours. He presented to the primary care clinic multiple times before the imaging was ordered. Subsequent MRI showed fibrous dysplasia of the right lesser wing of the sphenoid bone. After the surgery, the headache improved as well as his quality of life.

**Keywords:** *Headache, MRI, Bone Tumours, Sphenoid Bone Fibrous, Migraine*

## INTRODUCTION

Fibrous dysplasia is a benign primary disorder of bone of unknown aetiology. It is an uncommon disorder. It occurs due to a defect in osteoblastic differentiation and maturation which lead to abnormal bone development. Fibrous dysplasia involving the skull base most commonly involved ethmoid bone followed by sphenoid bone. There are 3 subtypes of fibrous dysplasia; Monostotic; fibrous dysplasia with single bone involvement only, which represent 70% of cases, Polyostotic; fibrous dysplasia involving multiple bones and McCune Albright syndrome; fibrous dysplasia of the bone associated with hyperthyroidism, sexual precocity and cafe au lait macular spot. (Singh & Mohan, 2007) This represents 3-5% of fibrous dysplasia patients.

## CASE PRESENTATION

A 31 years old gentleman, presented with a chief complaint of headache for 1 year. He described it as a generalized headache, worse on the right side and dull in nature. The headache occurred almost every day and can last throughout the day if analgesia was not used. He described the headache as moderate to severe intensity. He felt that his headache was getting worst compared to 1 year ago in terms of frequency as well as severity. The headache frequently woke him up during sleep. It was aggravated by coughing and straining. There was no relieving factor except for temporary relief with analgesics. It was associated with vomiting on and off as well as the right-sided blurring of vision. He reported that he almost had an accident two months ago due to his visual problem while on the way to his workplace, riding a motorcycle. There was no preceding aura, no photophobia, no limbs weakness, no fever, no neck stiffness, no runny nose, or watery eye. On further history, he has already been to our primary care clinic several times for the same complaint. It was noted that he had come to our clinic 6 times in the past year. Unfortunately, the problem was not resolved and neither did it improve. He was treated as a “migraine” and “tension-type headache”

by the doctors who saw him then. He was also started on migraine prophylaxis (propranolol 40mg BD) which did not reduce his headache despite his being compliant with the medication.

He was diagnosed with hypertension a few months ago. The diagnosis was made when he came for his complaint of headache, and it was noted that his blood pressure was persistently elevated at every visit. In fact, he said that some of the doctors who saw him at the previous visit told him that his headache is due to his high blood pressure. His blood pressure readings ranged during his previous visit between 140- 154/84-98mmHg. He was started on perindopril 8mg daily. He was also diagnosed with Diabetes Mellitus recently after two fasting glucose readings were 13.9mmol/L and 7.9mmol/L respectively. He, however, did not have any hyperglycaemic symptoms. He was started on Metformin 500mg twice daily 2 months ago.

On examination, his blood pressure was 144/90 with a regular heart rate of 80 bpm. He looks comfortable and did not appear anxious or depressed. He was afebrile. He was neither in pain nor respiratory distress. There was no gross deformity, or any abnormal movements seen. There was no facial asymmetry noted. His speech was normal with no slurred speech noted. There was no neurocutaneous stigmata present. His neurological examination specifically cranial nerve showed he has right unilateral temporal hemianopia. Other examinations were unremarkable. Subsequently, an MRI of the brain was done and showed the right lesser wing of the sphenoid bone is expanded with iso to hypodense lesion seen within. This mass expands into the right orbita l la teral wa ll with a mild displa cement of the la teral rectus muscle. He was then subsequently scheduled for an excisional biopsy which was done through a frontal craniotomy. The mass was taken out. The biopsy result confirmed the diagnosis of benign fibrous dysplasia of the right sphenoid bone.

**RESULTS**

MRI BRAIN	right lesser wing of the sphenoid bone is expanded with iso to hypodense lesion seen within. This mass expands into the right orbital lateral wall with a mild displacement of the lateral rectus muscle. No other lesions were noted. Impression; fibrous dysplasia of right lesser wing of the sphenoid bone. Differential; sclerotic metastases
-----------	--

After the surgery, he felt much better. He had no more headaches or blurred vision. Apart from that, his blood pressure also normalised. The doctor in the ward stopped his hypertension medication after surgery and his blood pressure reading come back to normal without medication.

**DISCUSSION**

Fibrous dyspla sia is a loca lised bone condition marked by aberrant fibrous tissue proliferation mixed with normal or immature bone, endocrine dysfunction, abnormal pigmentation, and early puberty in girls (Lustig et al., 2001). In cases of headache, neuralgia, sensory disturbance, functional issue, or infectious ENT complications, a diagnosis of craniofacial fibrous dysplasia should be suspected (Couturier et al., 2017). It's a benign bone tumour with the potential to cause serious cosmetic and functional problems, especially in the craniofacial skeleton. The surgeon faces major difficulties in managing it. Its compression of the optic nerve, which results in visual impairment, is particularly concerning (Chen, Chang, & Tan, 2006).

As in this patient, the main issue is about the missed diagnosis which occurred for quite a long time. The first history taking gathered from the patient himself, as well as from the previous visit record showed that he had come for a similar headache problem so many times within 6 months. Unfortunately for him, no one was able to at least suspect his headache is secondary to a space-occupying lesion inside his brain. In order to avoid this mistake, it is always about good history taking and proper physical examination. From his records, none were mentioned about the visual field and funduscopy findings as well as the related cranial nerve examination. In fact, in most of the documentation, the history taking was very brief without mentioning the red flag symptoms which this patient had; abnormal neurological examination, the headache that awakened the patient from sleep and headache precipitated by physical exertion or Valsa lva manoeuvre (eg coughing,

laughing, straining). Having these 3 red flag symptoms are enough to suspect secondary headache rather than just migraine/ tension-type headache.

Having the fact that this patient has had a chronic headache for more than 6 months should have alerted the primary care doctors to use headache assessment tools namely headache diary. The use of a headache diary has been proven to improve the diagnosis. The headache diary is important to record the patient's symptoms including the severity and frequency of the headache. It is also used to assess the effectiveness of the intervention which is important as it will give us a clue whether we are treating the patient correctly. Failure to improve with the intervention given may suggest the need to revise our diagnosis, just like for this patient (Suetterlin, & Turner, 2014; Russell et al., 1991; Phillip, Lyngberg & Jensen, 2007). Having a chronic headache for more than 6 months should have alerted the primary care doctors to use headache assessment tools namely headache diary. The use of a headache diary has been proven to improve the diagnosis. It is important to record the patient's symptoms including the severity and frequency of the headache (Ooi, 2018; Saleem, Javed, & Ahmad, 2020; Harris, Poddar, Gitelis, Sheinkop & Rosenberg, 1995).

## CONCLUSION

Headache is a common condition that has significant social morbidity. In order to determine the underlying aetiology and make a positive diagnosis, a thorough history must be obtained. This is crucial to reduce long-term morbidity, avoid mortality, and enhance patient outcomes.

## Conflict of interest

The authors declare that they have no competing interests in writing this article.

## ACKNOWLEDGEMENT

The authors would like to thank the patient for his permission and cooperation in writing this case report. Informed consent of the case was obtained before the preparation of the case report.

## REFERENCES

- Chen, Y., Chang, C., & Tan, Y. (2006). Craniofacial fibrous dysplasia: an update. *Chang Gung medical journal*, 29(6), 543.
- Couturier, A., Aumaître, O., Gilain, L., Jean, B., Mom, T., & André, M. (2017). Craniofacial fibrous dysplasia: a 10-case series. *European annals of otorhinolaryngology, head and neck diseases*, 134(4), 229-235. <https://doi.org/10.1016/j.anorl.2017.02.004>
- Lustig, L. R., Holliday, M. J., McCarthy, E. F., & Nager, G. T. (2001). Fibrous dysplasia involving the skull base and temporal bone. *Archives of otolaryngology–head & neck surgery*, 127(10), 1239-1247. [10.1001/archotol.127.10.1239](https://doi.org/10.1001/archotol.127.10.1239)
- Phillip, D., Lyngberg, A. C., & Jensen, R. (2007). Assessment of headache diagnosis. A comparative population study of a clinical interview with a diagnostic headache diary. *Cephalalgia*, 27(1), 1-8. [10.1111/j.1468-2982.2007.01239.x](https://doi.org/10.1111/j.1468-2982.2007.01239.x)

Russell, M. B., Rasmussen, B. K., Brennum, J., Iversen, H. K., Jensen, R. A., & Olesen, J. (1992). Presentation of a new instrument: the diagnostic headache diary. *Cephalalgia*, 12(6), 369-374. [10.1111/j.1468-2982.1992.00369.x](https://doi.org/10.1111/j.1468-2982.1992.00369.x)

Singh, H., & Mohan, C. (2007). Fibrous dysplasia of Sphenoid bone. *Indian Journal of Otolaryngology and Head & Neck Surgery*, 59(2), 157-159. [10.1007/s12070-007-0046-x](https://doi.org/10.1007/s12070-007-0046-x)

Suetterlin, K. & Turner, C. (2014). Diagnosis and management of headache. *British Journal of Hospital Medicine*, 75(Sup12), pp.C178-C182. [10.12968/hmed.2014.75.Sup12.C178](https://doi.org/10.12968/hmed.2014.75.Sup12.C178)

Ooi, F. K. (2018). BONE PROPERTIES AND BLOOD BONE METABOLISM MARKERS IN RESPONSE TO EXERCISES AND HONEY SUPPLEMENTATION: A CONTEMPORARY REVIEW. *Malaysian Journal of Medical Research (MJMR)*, 2(2), 68-74. <https://doi.org/10.31674/mjmr.2018.v02i02.010>

Saleem, S., Javed, F., & Ahmad, I. (2020). MANAGEMENT OF LARGE SIZE AND MULTIPLE BENIGN BREAST LESIONS USING ONCOPLASTIC SURGICAL SKILLS. *Malaysian Journal of Medical Research (MJMR)*, 4(3), 6-10. <https://doi.org/10.31674/mjmr.2020.v04i03.002>

Harris, A. I., Poddar, S., Gitelis, S., Sheinkop, M. B., & Rosenberg, A. G. (1995). Arthroplasty with a composite of an allograft and a prosthesis for knees with severe deficiency of bone. *The Journal of Bone and Joint surgery. American Volume*, 77(3), 373-386. [10.2106/00004623-199503000-00007](https://doi.org/10.2106/00004623-199503000-00007)