CASE REPORT

Multiple lesions of skull and cervical spine: a rare presentation of unicameral bone cysts

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SUMMARY

A 55-year-old man with a history of Benign Paroxysmal Positional Vertigo unalleviated by Epley manoeuvre presented to an otolaryngologist for dizziness, right ear fullness and headache. MRI of the brain showed numerous marrow-replacing lesions throughout the calvarium, skull base and upper cervical spine which were hypointense on T1-weighted images, hyperintense on T2-weighted images and avidly enhanced following contrast, concerning for a malignant process such as metastatic disease or multiple myeloma (figure 1). Systemic X-ray survey (spine, skull, chest, pelvis, all long bones) and nuclear medicine whole body bone scan were negative except for the lesions seen on MRI. β -2microglobulin, immunoglobulin and monoclonal protein electrophoresis were negative for myeloma or immunological process. Given the concern for metastatic disease, biopsy of a skull lesion was recommended. Pathological analysis of a calvarial lesion was consistent with unicameral bone cyst (figure 1). No ongoing therapy was offered; however, brain and spine surveillance imaging will continue.

BACKGROUND

Unicameral bone cysts (UBCs) are benign, fluid-filled cavities that develop primarily in tubular and flat bones (eg, humerus, femur) but can affect any bone. These lesions usually expand and weaken the surrounding bone; however, they are not true neoplasms. First described in 1876 by Virchow as cystic structures caused by abnormalities in local circulation,¹ UBCs occur almost exclusively in children and adolescents (up to 85% of cases).^{2,3} These lesions represent approximately 3% of all bone tumours and have a male predominance.¹

UBC has been reported in the skull/calvarium; however, the majority of cases were solitary lesions and were found in adolescents.⁴⁻⁶ The aetiology of UBCs remains unclear; however, rare associations with severe bilateral malformations of the ear, choanal atresia and neurofibromatosis have led some to believe that there may be a congenital component.⁵ To our knowledge, this is the first report of UBC in an adult with numerous skull and spine lesions. Interestingly, the patient's father also had a skull lesion of unknown aetiology diagnosed more than 10 years before his death.

CASE PRESENTATION

Presenting features: Intermittent dizziness and vertigo with intermittent right ear fullness and tinnitus, headache.

Medical history: No medical history.

Social history: Never smoked, drinks one time or more per month, two cups of coffee per day.

Family history: Hearing loss (mother and father), cancer (father), heart disease (father), diabetes (mother).

INVESTIGATIONS

- ▶ MRI of the brain (Figure 1).
- ▶ X-ray skeletal survey (skull, chest, pelvis, spine, extremity long bones) (Figure 1).
- ▶ CT Head (Figure 1)
- ▶ β -2microglobulin, immunoglobulin and monoclonal protein electrophoresis.
- ▶ Nuclear medicine whole body bone scan.
- ▶ Biopsy of a skull lesion.

DIFFERENTIAL DIAGNOSIS

- ▶ Metastatic disease.
- ▶ Myeloma.
- ▶ Aneurysmal bone cyst.
- ▶ Epidermoid/Acquired cholesteatoma.
- ▶ Cholesterol granuloma.
- ▶ Local manifestation of histiocytosis X (Eosinophilic granuloma).
- ▶ Granulomatous process.
- ▶ Fibrous dysplasia.
- ▶ Osteitis and osteomyelitis.
- ▶ Osteoporosis.
- ▶ Paget's disease.

Skull normal variants.

- ▶ Venous Lakes
- ▶ Emissary veins, transcranial
- ▶ Arachnoid granulations
- ▶ Prominent convolutional markings
- ▶ Parietal foramina

TREATMENT

No treatment is needed.

OUTCOME AND FOLLOW-UP

Follow-Up: Continue surveillance Brain MRIs.

DISCUSSION

A survey of the literature reveals some reports of UBCs in the skull, but most of these are in the temporal bone and nearly all of those found in the skull were in patients in their first or second decade of life.⁵⁻⁷ The aetiology is unclear; however, unicameral/solitary bone cysts have been associated with severe bilateral malformation of the ear, choanal atresia and neurofibromatosis,^{8,9} suggesting a possible congenital or genetic origin.

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Figure 1 (A) X-ray skull shows lucencies throughout the calvarium consistent with lytic lesions. (B and C) Sagittal and axial head CT reconstruction and axial head CT demonstrate numerous lytic lesions throughout the calvarium and cervical spine which correspond to the areas of contrast enhancement seen on MRI. (D) Sagittal precontrast MRI. (E) Sagittal postcontrast MRI (same cut as (D)) shows numerous mostly tiny enhancing lesions throughout the skull. (F) Axial postcontrast MRI, cut showing C3 lesion. (G) Axial post contrast MRI further demonstrating numerous, small, avidly enhancing skull lesions, and midline sagittal postcontrast MRI shows not only the skull lesions, but also the enhancing cervical spine lesion which nearly completely replaces the fatty signal within the C3 vertebral body. (H) Section of a portion of the UBC wall and overlying calvarium. The membrane lining the inner surface of the cyst (partial/not intact) consists of a moderately dense fibrous connective tissue with occasional cuboidal endothelial cells (H&E stain 10×).

Patient's perspective

Some details of case presentation

- ▶ Ear fullness had been present for at least 2–3 years prior to dizziness/vertigo; has been largely mitigated by antihistamines (fexofenadine and budesonide nasal spray).
- ▶ Headache has not been frequent or severe. I have had occasional sinus headaches for 35 years, but about 7 months prior to lesion biopsy, began to experience a new type of headache. This new type of headache is localised to a very small (perhaps 1-inch diameter area and seems to emanate from between the skull and scalp. The headache sensation resembles an intense (but not especially painful) tingling and is of short (5–10 min) duration.

Details about father's skull lesion

At age 62, my father had an X-ray that produced an initial diagnosis of a bone tumour in the base of the skull. He sought a second opinion from Cleveland Clinic, which diagnosed his condition as osteoarthritis and not cancer. I am not certain if Cleveland Clinic had performed additional imaging, or simply reinterpreted the X-ray. He lived an additional 10 years before he died of a heart attack. I do not know what symptoms prompted the initial X-ray, and I do not recall him complaining of pain or other symptoms at the base of his skull.

General observation

It remains to be determined how or if the unicameral bone cysts may be related to the symptoms of dizziness and vertigo. A vestibular evaluation, using caloric irrigation, had found hypofunction of the right ear vestibular system. I do not know if any of the unicameral bone cysts are proximal to the right vestibular region. The ear fullness and dizziness seem to be exacerbated by activities (golfing and stretching) that involve holding the head still and rotating the shoulders or rotating the head side-to-side while holding the shoulders still.

Learning points

- ▶ Unicameral bone cysts (UBCs) can be multifocal.
- ▶ These lesions may occur in adults or present later life.
- ▶ UBCs of the skull/calvarium are rare but should be considered in the differential diagnosis of patients with lytic skull lesions.
- ▶ UBCs may have a congenital or genetic component.

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