Unusual presentation of more common disease/injury

Intracranial chondroma: a rare entity

Veena Maheshwari,¹ Ghazala Mehdi,¹ Manoranjan Varshney,¹ Anshu Jain,¹ Sonal Vashishtha,¹ Kavita Gaur,¹ Vinod Kumar Srivastava²

¹Department of Pathology, J.N. Medical College, AMU, Aligarh, Uttar Pradesh, India;

²Department of Surgery, J.N. Medical College, AMU, Aligarh, Uttar Pradesh, India

Correspondence to Kavita Gaur, kavgaur@gmail.com

Summary

Intracranial chondroma is a rare benign cartilaginous tumour with an incidence of less than 1% of all primary intracranial tumours. The authors are reporting here a case of intracranial chondroma in a 40-year-old man who presented with 5-month history of headache and gradual diminution of vision. A tentative diagnosis of chondroma was made on imprint cytology which was confirmed on histopathological examination.

BACKGROUND

Intracranial chondroma is a rare tumour with an incidence of 0.2–0.3% of all the intracranial tumours.

CASE PRESENTATION

A 40-year-old man presented with 5-month history of headache in the left frontal region with gradual diminution of vision. General and systemic examinations were unremarkable. Plain x-ray skull revealed a large tumour in the anterior cranial fossa (figure 1). CT scan showed an oval-shaped extra-axial mass in the left frontal region. The mass

was surgically removed and submitted for histopathological examination. Grossly, the tumour was found to be 8 cm in diameter, firm with a pseudocapsule (egg shell-like). Cut surface revealed a soft and greyish white material. Imprint cytology showed widely separated chondrocytes in a background of abundant bluish, glassy looking chondroid material (figure 2). A tentative diagnosis of chondroma was made.

Microscopic examination of the tumour was composed of hyaline cartilage which was partially lobulated by fibrous septa. The cellularity varied in different fields, and

Figure 1 X-ray skull (lateral view): large tumour in anterior cranial fossa expanding and partially destroying the frontal bone.

the chondrocytes occurred either singly or were arranged in pairs or tetrads. They were of various shapes and sizes and contained a single darkly stained nucleus. Nuclear pleomorphism, mitosis, high cellularity and myxoid fibrillary matrix were not seen (figure 3). On the basis of above findings, a final diagnosis of intracranial chondroma was rendered.

DIFFERENTIAL DIAGNOSIS

- Meningioma
- chondrosarcoma.

TREATMENT

Surgical excision of tumour was done.

Figure 2 Imprint cytology: scattered chondrocytes in a background of pale bluish chondroid material (H&E $\times 250$).

Figure 3 Histopathological picture: chondrocytes in pairs with single darkly stained nuclei in a background of hyaline cartilage (H&E $\times 125$).

OUTCOME AND FOLLOW-UP

Patient was discharged in good conditions.

DISCUSSION

Chondromas are benign tumours which can be located in any part of the body. Intracranial chondroma is a benign cartilaginous tumour. These may be solitary and when multiple form a part of Ollier's disease¹ or Maffucci's syndrome.² Studies shows that these tumours usually arise from the skull base,^{3–4} although they may rarely occur from other parts of the intracranial cavity.^{3–5–6}

It comprises 0.2–0.3% of all intracranial neoplasms.^{7–8} They were first described by Hirshfield in 1851 and the first operative resection was published by Nixon in 1982. A majority of these tumours are extradural and generally arise from the skull base. Data claim only 15–30% tumours to be intradural.^{9–10} The origin of intracranial chondroma is still debatable. Several theories have been put forward explaining the preponderance of these chondromas originating in the skull base. Tumours arising from dura, choroid plexus, leptomeninges or within brain parenchyma are less common. The chondromas arising from dura are totally ensheathed by layers of the dura or attached to the subdural surface of the dura or falx. The most accepted one states their origin from the embryonic rests of cartilage cells at the basilar synchondroses^{5–11} whereas the intradural origin is better explained by metaplasia of meningeal fibroblasts or perivascular mesenchymal tissue. Some authors have also proposed other origins like ectopic embryologic rest of cartilage cells or traumatic displacement of cartilaginous elements. There are very few intraparenchymal chondromas. Out of these, only a few are truly parenchymal and arise by metaplasia in an existing glioma.^{5–12}

Some of the skull base tumours have been reported as a component of Ollier's disease as well as Maffucci's syndrome. Malignant change has also been reported in a case of intracranial chondroma with Maffucci's syndrome.¹³

The intracranial chondromas generally occur in the third decade of life,^{9–14} although cases have been reported in 15 months to 60 years of age and no preference to either sex.¹⁴

Zulch¹⁵ found that these chondromas behave in a similar fashion to their counterparts found elsewhere in the body except that they have an irregular pattern and grow predominantly by expansion. Clinically, intracranial chondromas may present with headache, seizures or focal neurological deficit depending upon their site.¹⁰ Because the intradural chondromas are generally slow growing, patients usually present with a long history of clinical symptoms and with large masses at the time of diagnosis. Chondromas are avascular structures which might compress but do not invade the brain substance.

Grossly, these tumours may present as firm, lobulated, greyish white masses with or without calcification. Microscopically, the tumour is well circumscribed and has a fine fibrous capsule. It is composed of single or paired chondrocytes in lacunae with a background of abundant chondroid matrix. The chondromas form hyaline cartilage which may be trabeculated and lobulated. No mitoses or nuclear pleomorphism is generally seen.

The diagnosis can be confirmed preoperatively with the help of CT scan, MRI and even plain x-ray skull. On x-ray,

they may present with hyperostosis especially of the inner table of skull which may indicate bone destruction occurring in up to 50% of intracranial chondromas. Sometimes there may be areas of calcification. On CT scan, it appears as a well circumscribed, extra-axial, avascular mass^{5–9} with globular or diffuse calcification which may demonstrate slight enhancement after contrast injection.

Meningioma and chondrosarcoma form the differential diagnosis of the intracranial chondroma.¹⁶ Meningioma may mimic chondroma clinically but radiologically it can be clearly differentiated. Its cartilaginous variant can be differentiated histologically by the presence of foci of cells with meningothelial features.¹⁷ Microscopically, chondrosarcoma can be differentiated by the presence of nuclear pleomorphism and mitotic activity, but sometimes very well differentiated cartilaginous areas can be found within a chondrosarcoma. This emphasises the need for careful pathological evaluation of the specimen before labelling it as benign chondroma.

The prognosis of intracranial chondroma is fairly good. Surgical removal remains the treatment of choice.⁹ If total removal cannot be done, partial removal along with the decompression of neural structure proves to be a beneficial modality of treatment.

Learning points

- ▶ Chondroma can be seen in any organ but its occurrence in intracranial cavity is rare.
- ▶ They have malignant potential when associated with Maffucci's syndrome.
- ▶ Intraparenchymal tumour is extremely rare.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Trafflet RF**, Babaria AR, Barolat G, *et al.* Intracranial chondroma in a patient with Ollier's disease. Case report. *J Neurosurg* 1989;**70**:274–6.
2. **Chakraborty S**, Tamaki N, Kondoh T, *et al.* Maffucci's syndrome associated with intracranial enchondroma and aneurysm: case report. *Surg Neurol* 1991;**36**:216–20.
3. **Burger PC**, Scheithauer BW, Vogel FS. *Surgical Pathology of the Nervous System and Its Coverings*. Third edition. New York, NY: Churchill Livingstone 1991:96–8.
4. **Burger PC**, Scheithauer BW. *Tumors of the Central Nervous System: Atlas of Tumor Pathology*. Third series, fascicle 10. Washington, DC: Armed Forces Institute of Pathology 1994:302.
5. **Ahyai A**, Spoerri O. Intracerebral chondroma. *Surg Neurol* 1979;**11**:431–3.
6. **Chorobski J**, Jarzyski J, Ferens E. Intracranial solitary chondroma. *Surg Gynecol Obstet* 1939;**68**:677–86.
7. **Colpan E**, Attar A, Ereku S, *et al.* Convexity dural chondroma: a case report and review of the literature. *J Clin Neurosci* 2003;**10**:106–8.
8. **Nakayama M**, Nagayama T, Hirano H, *et al.* Giant chondroma arising from the dura mater of the convexity. Case report and review of the literature. *J Neurosurg* 2001;**94**:331–4.
9. **Mapstone TB**, Wongmongkolrit T, Roessman U, *et al.* Intradural chondroma: a case report and review of the literature. *Neurosurgery* 1983;**12**:111–14.
10. **Sebbag M**, Schmidt V, Leboucq N, *et al.* [Dura mater chondroma. A case report and review of the literature]. *J Radiol* 1990;**71**:495–8.
11. **Acampora S**, Troisi F, Fusco G, *et al.* Voluminous intracranial chondroma. *Surg Neurol* 1982;**18**:254–7.
12. **Hirvonen J**, Heikinheimo H. A case of intracerebral chondroma. A case report. *Acta Pathol Microbiol Scand* 1969;**76**:19–24.

13. **van Nienlen KM**, de Jong BM. A case of Ollier's disease associated with two intracerebral low-grade gliomas. *Clin Neural Neurosurg* 1999;**101**:106–10.
14. **Dutton J**. Intracranial solitary chondroma. Case report. *J Neurosurg* 1978;**49**:460–3.
15. **Zulch KJ**. *Brain Tumours*. Third edition. New York, NY: Springer-Verlag 1986:479–81.
16. **Higashida T**, Sakata K, Kanno H, *et al*. [Intracranial chondroma arising from the skull base: two case reports featuring the image findings for differential diagnosis]. *No Shinkei Geka* 2007;**35**:495–501.
17. **Russell DS**, Rubinstein LJ. *Pathology of Tumours of the Nervous System*. Fifth edition. London: Edward Arnold 1989.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Maheshwari V, Mehdi G, Varshney M, Jain A, Vashishtha S, Gaur K, Srivastava VK. Intracranial chondroma: a rare entity. *BMJ Case Reports* 2011; 10.1136/bcr.03.2011.4047, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow