A Unique Presentation of Retroclival Chordoma

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Abstract:
Chordomas are rare tumours which arise from remnants of the primitive notochord. They occur primarily in the sacrum, clivus and cervical regions. We report a case of retroclival chordoma which presented as an extradural haemorrhage following minor trauma. The underlying tumour was not apparent on imaging performed immediately following the event, and chordoma presenting in this manner has not previously been described in the literature. The tumour became apparent on subsequent imaging, and progressed despite surgical debulking and radiotherapy. (J Postgrad Med 2002;48:285-287)

Key Words: Chordoma, haemorrhage, Magnetic resonance imaging

Retroclival chordomas are rare tumours, accounting for approximately 1% of all intracranial tumours. They usually present with cranial nerve palsies, visual disturbances and orbito-frontal headaches. They rarely have intracerebral haemorrhage as the presenting feature. We report on a patient with retroclival chordoma, whose first presentation was with an extradural haemorrhage following minor trauma. This clinical and radiological presentation of retroclival chordoma has not previously been reported.

Case History
A 29-year-old man presented with an eight day history of headache after a low-speed road traffic accident, where he was shunted from behind whilst driving his car at approximately 30mph. On examination, his Glasgow Coma score was 15/15, and he had bilateral cranial nerve VI palsies and an absent gag reflex.

A CT scan (Figure 1) showed an extradural haematoma compressing and distorting the brainstem. Subsequent MRI (Figure 2) confirmed the presence of a subacute extradural haematoma anterior to the brainstem. No source of bleeding was identified.

The patient’s cranial nerve palsies progressed, and he developed diplopia. He then underwent diagnostic cerebral angiography. This study proved difficult to interpret due to the mass effect from the haematoma, but no definite vascular malformation was identified.

The patient then underwent repeat MRI scanning one month after the initial presentation. This scan identified a mass lesion at the upper aspect of the subacute extradural haematoma (Figure 3). The mass was well-defined, slightly lobulated and showed pathological enhancement following the administration of gadolinium-DTPA. He then went on to have a craniotomy for evacuation of the haematoma with biopsy and debulking of the tumour mass. The histology (Figure 4) showed tumour cells in sheets and cords within a mucoid background. The cells were uniformly cuboidal with some large phsalliphorous cells with foamy cytoplasm. The findings were consistent with a diagnosis of chordoma.

His symptoms and neurological deficit improved following surgery, and he went on to have a course of proton beam radiotherapy. However, a follow-up MRI scan (Figure 5) showed progression of the lesion despite treatment.

Discussion
Chordomas are malignant neoplasms derived from embryonic notochordal remnants. While the majority of chordomas occur in the sacral region, more than one third arise in the skull base. As the terminus of the notochord is in the sphenoid bone inferior to the sella turcica, skull base chordomas arise adjacent to the clivus.1 These tumours usually produce marked cranial nerve palsies, visual disturbances and orbito-frontal headaches.

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Figure 1: Axial unenhanced CT scan demonstrates an extradural haematoma which compresses and distorts the brainstem.
Figure 2: Sagittal T1-weighted MRI scan confirms the presence of a subacute extradural haematoma.

Figure 3: MRI scans performed one month after the initial presentation. The sagittal T1-weighted image demonstrates a mass lesion at the superior aspect of the haematoma.

Figure 4: Histological specimen shows the typical appearance of a chordoma.

Figure 5: Axial T2-weighted scan reveals progressive disease despite surgical debulking and radiotherapy. The image shows areas of low signal within the lesion, consistent with haemorrhage and/or calcification.

destruction of the sphenoid bone and extend into the sphenoid sinus and nasopharynx.

Cranial nerve palsies due to tumour extension into the neural foramina is the most frequent presentation of clivus chordoma. There may be associated headache, signs and symptoms of raised intracranial pressure and pyramidal signs. Acute presentation following intraparenchymal haemorrhage has also been described.

The CT appearance of a clivus chordoma is usually a midline lesion with bone destruction and a soft-tissue mass. The bone adjacent to the tumour does not have a sclerotic margin, and large calcific fragments are frequently seen. There is a variable appearance on MRI, with a hypointense to isointense soft-tissue mass seen on T1-weighted images and high signal on T2-weighted scans. Cystic areas containing haemorrhage are frequently present, and produce high signal on T1-weighted images.

Proton beam radiotherapy following surgical debulking is the preferred treatment of these tumours, which are rarely amenable to complete resection. This combined treatment can achieve 3-year survival rates of up to 91%.

Chordomas are frequently haemorrhagic, and can rarely present as a primary intracerebral haemorrhage. However, the underlying tumour has been demonstrated on imaging at presentation in all previously reported cases. Chordoma presenting as an extr-
tradural haemorrhage following trauma and remaining occult on initial imaging has not previously been reported. This case demonstrates the importance of seeking an underlying cause for an extradural haematoma which occurs at an unusual site following relatively minor trauma.

References

The diagnosis is Osler-Weber-Rendu syndrome.

Osler-Weber-Rendu syndrome, also known as hereditary haemorrhagic telangiectasia (HHT), is an autosomal dominant mucocutaneous and visceral fibrovascular dysplasia that is usually recognised as a “classic triad” of telangiectasia, recurrent epistaxis and a family history of the disorder. Most patients with HHT report similarly affected relatives. However in 20% of cases, there is no family history of either telangiectasia or recurrent bleeding. Such cases could possibly be due to sporadic spontaneous mutation. This disorder occurs in all races with an estimated frequency of one or two per 100,000. The vascular dysplasia of HHT is localised to discrete segments of vessels ranging in caliber from capillaries to large arteries and veins. It results in three forms of dysplasia-telangiectasia, arteriovenous malformations and aneurysms. The characteristic lesion of this syndrome is the macular telangiectasia, a punctiform spot, 1 to 3 mm in diameter, most commonly over the face, followed by lips, nares, tongue, ears, hands, chest and feet, often increasing in size and number with age. Ninety five percent of patients have had nosebleeds by 20 years. Visible telangiectasia appear on the skin and mucous membrane 5 to 20 years after epistaxis begins and the disease usually declares itself by the age of 40 years. Gastrointestinal bleeding develops in the 4th to 5th decade. Other features include pulmonary arteriovenous fistulas (clinically manifesting as clubbing, cyanosis and polycythemia), retinal telangiectasia, hepatic fibrotic arteriovenous malformations, central nervous system aneurysms, arteriovenous malformations, and telangiectases. Epistaxis and melaena are the frequent complications of HHT due to telangiectasia in the gastrointestinal tract and nasal mucosa. Other syndromes with telangiectasia include systemic scleroderma (CREST syndrome), generalised essential telangiectasia (occurs in women in their 4th and 5th decades, begins on the legs and slowly involves thighs and lower abdomen) and ataxia telangiectasia (presents as cerebellar ataxia in children shortly after they begin to walk). Our case had telangiectasia, epistaxis, and haematuria, and thus represents a typical example of Osler-Weber-Rendu syndrome.

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