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January 2010

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## Recommended Citation

Jumani, M., Channa, R., Zafar, S., Enam, S. (2010). Pontine hemorrhage due to chondrosarcoma of the skull base. *Journal of College of Physicians and Surgeons Pakistan*, 20(10), 692-694.

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# Pontine Haemorrhage Due to Chondrosarcoma of the Skull Base

Maliha Iqbal Jumani, Roomasa Channa, Sayed Nabeel Zafar and Syed Ather Enam

## ABSTRACT

A young adult presented with acute weakness of right side of body and slurring of speech. An initial brain CT scan showed a pontine haemorrhage, however MRI done a few days later revealed a mass in the left parapharyngeal space. Histopathology of the mass revealed that it was a chondrosarcoma. Subsequently the patient was initially treated with chemotherapy and radiotherapy and later surgery. Skull base chondrosarcoma is to be included in the differentials of a young patient presenting with signs consistent with pontine haemorrhage.

**Key words:** *Chondrosarcoma. Pontine haemorrhage. Skull base.*

## INTRODUCTION

Pontine haemorrhage represents approximately 6% of intracranial haemorrhages.<sup>1</sup> The common causes of pontine haemorrhage are arterial hypertension, vascular malformations and anti-coagulant therapy.<sup>2</sup> Other causes include inflammatory vascular disease, cerebral amyloid angiopathy, head trauma and intracranial neoplasms. Pontine haemorrhage due to a tumour has rarely been described.<sup>3</sup> Tumours, however, do cause haemorrhage in the brain elsewhere. Intratumoural haemorrhage often occurs in gliomas, most commonly glioblastomas or oligodendrogliomas. Metastatic tumours to the brain that are most likely to bleed include melanomas, thyroid carcinomas, renal cell carcinomas and choriocarcinomas.<sup>4</sup> Extra-axial tumours that may present with brain haemorrhage include meningiomas, acoustic neuromas and chordomas. Chondrosarcoma is an extra-axial tumour that has been reported to cause intracranial haemorrhage.<sup>5</sup>

However, to the best of our knowledge chondrosarcomas causing pontine haemorrhage have not been reported as yet. Chondrosarcomas are malignant tumours composed of chondrocytes and are rarely seen intracranially. Here this report describes the case of a young man presenting with pontine haemorrhage, later diagnosed to be due to chondrosarcoma.

## CASE REPORT

A 21-year-old man presented to the emergency department with a 2 hours history of weakness on the right side of body, slurring of speech and vomiting. There was no history of headaches, fits or unconsciousness.

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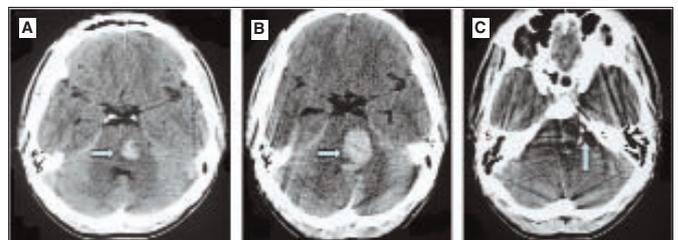
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Received May 11, 2009; accepted August 19, 2010.

On general physical examination, the patient was afebrile and had a blood pressure of 125/78 mmHg, pulse of 96/minute and a respiratory rate of 14/minute. The patient's GCS at the time of arrival was 14/15.

Examination of the cranial nerves showed mildly dilated left pupil which was sluggish to light, ptosis and horizontal nystagmus of the left eye, left facial weakness and right hemiparesis. Examination of the cardiovascular system, chest and abdomen were within normal limits. A brain CT scan of the patient at the time of admission showed a pontine haemorrhage measuring 1.8 x 1.9 cm associated with minimal surrounding oedema (Figure 1). During the first 24 hours of hospital admission, the patient's GCS deteriorated to 8/15. His respiratory effort decreased and he had multiple episodes of vomiting and aspiration. He was intubated and kept on ventilatory support. A repeat CT scan at that time showed an increase in the size of the haemorrhage to 2 x 2.5 cm, and an additional finding of a mass in the skull base, a feature that was previously missed.



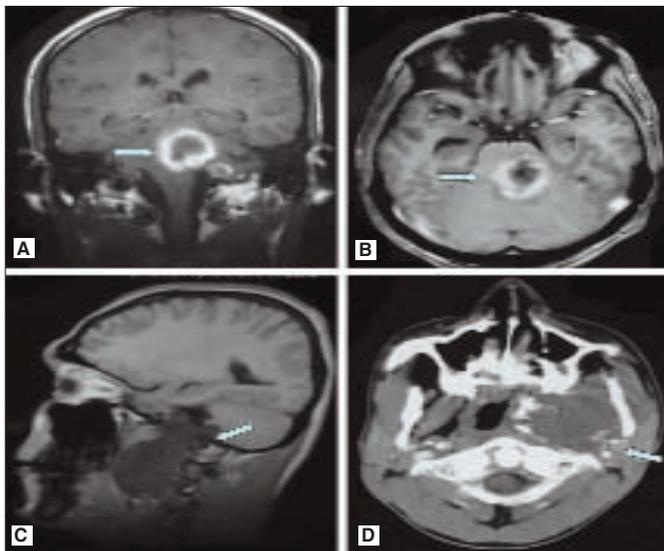
**Figure 1:** Axial CT scans of the brain.

**A:** At the time of presentation, showing pontine haemorrhage with minimal surrounding edema (left arrow).

**B:** Repeated a few hours later on the same day showing an increase in size of the haemorrhage (centre arrow).

**C:** Specks of calcification (right arrow) seen in the posterior cranial fossa at the level of the petrous apex.

A brain MRI, done a few days later with the patient medically stable, supported the findings of the last CT scan (Figure 2). It showed a mass in the left parapharyngeal space eroding into the oropharynx and extending into the cranial cavity through the petrous bone. The mass was also reported as eroding into the brainstem and compressing the fourth ventricle.



**Figure 2:** Magnetic resonance images of the brain done a week after admission.  
**A:** Coronal section with Gadolinium showing area of contrast enhancement at the level of pons.  
**B:** Axial section with Gadolinium showing similar enhancement.  
**C:** T1 weighted sagittal image showing a hypointense and homogeneous mass eroding through the petrous apex into the brainstem.  
**D:** Axial section CT scan shows hypodense mass (arrow) with speckles of hyperdensity suggesting calcification as well as bony erosion at the base of the skull.

Subsequently a transnasal biopsy of the lesion was taken. The histopathology report showed tissue with predominantly cartilaginous differentiation and bony fragments exhibiting central fatty marrow spaces. Small pleomorphic cells with hyperchromatic nuclei separated by chondromyxoid matrix were seen. Immunostaining was positive for vimentin and S-100 but negative for cytokeratin CAM 5.2 and EMA. Based on these findings, the tumour was identified as an undifferentiated neoplasm with extensive chondroid differentiation, most likely a chondrosarcoma.

The proximity of the tumour to the brainstem called for the conservative methods of radiotherapy and chemotherapy as the initial treatment. Gross total resection of the tumour was undertaken later, when the tumour did not respond to three cycles of chemotherapy and radiotherapy. Surgery was performed using a combined and staged approach by a Neurosurgeon and an ENT surgeon. The patient was discharged with stable vitals and a GCS of 15/15 but with right hemiparesis (a deficit that remained since his first presentation to the hospital three months back). Five days postoperatively he presented to the ER with meningitis and hydrocephalus. He was hypotensive with loss of brainstem reflexes. Resuscitation with inotropic agents and ventilatory support was tried for 2 days but eventually withdrawn as no progress was observed. The cause of the patient's demise was speculated as meningitis due to oral pathogens gaining access to the central nervous system through the surgical incision that was not healing well. Wound dehiscence was attributed to the pre-operative radiation therapy given to the patient.

## DISCUSSION

This young gentleman presented with pontine haemorrhage due to intracranial chondrosarcoma. Chondrosarcomas are malignant tumours composed of chondrocytes. They represent 0.15% of all intracranial tumours. There are three types: classic or skeletal subdivided into three histological grades of increasing malignancy with evident chondroblastic differentiation and areas of hyaline cartilage matrix; mesenchymal - the most frequent aggressive variant at this location which histologically resembles primitive cartilaginous mesenchyme; myxoid - the rarest variant which lacks hyaline cartilage areas and has a prominent myxoid matrix.<sup>6</sup> Based on the histopathology report, the patient was diagnosed with low-grade skeletal chondrosarcoma.

Low grade chondrosarcomas are known to be invasive indolent tumours.<sup>7</sup> The case presented here highlights the occasionally highly invasive nature of this tumour. The tumour eroded through the bone, dura and blood vessels leading to haemorrhage at the time of presentation. Even though chondrosarcomas have been reported in literature as invasive, only 2 cases leading to haemorrhage have been recorded.<sup>5</sup>

This patient presented with multiple cranial nerves deficits. The signs and symptoms at the first manifestation of tumour are usually caused by oculomotor dysfunction as the tumour mainly arises from the petrous bone close to the midline.<sup>8</sup> Unilateral ptosis and pupillary dilation seen in this patient was consistent with this finding, but was most likely due to compression by the pontine haemorrhage rather than direct tumour invasion of the cranial nerves. Chondrosarcomas are generally thought of as slow growing tumours,<sup>9</sup> with a gradual onset of vague symptoms. Therefore, the short history of presenting symptoms in this case is the feature that is most striking. The short disease course in the patient along with the pontine haemorrhage that masked the tumour on the first CT scan, caused the primary team to overlook chondrosarcoma as a possible differential in the case. It was only after obtaining the histopathology report that a confirmed diagnosis of chondrosarcoma was made.

Previous literature published in this regard shows that it generally does take a long time to diagnose chondrosarcomas and a review of 177 cases showed that the mean duration of symptoms before diagnosis was 27 months.<sup>8</sup> In this case, the severity and acuteness of symptoms called for immediate intervention and therefore, all efforts were made to expedite the diagnosis which was done within a week. Several treatment options are available to manage patients with a chondrosarcoma. Most cases that have been reported in literature were treated with neurosurgery and conventional or proton radiotherapy. In 80% cases resection

was sub-total.<sup>8</sup> In a study of 15 patients, 13 of whom were operated on, 2 received proton therapy, and 01 received radiation therapy postoperatively, recurrence free survival rates at 2, 3, and 5 years intervals were 67%, 56% and 43% respectively.<sup>8</sup> In another study on 60 patients with chondrosarcoma, recurrence free survival rate at 5 years was 65%.<sup>8</sup> Literature recommends an optimal tumour resection in one go as the best treatment for intracranial chondrosarcomas as repeated surgical intervention is associated with risks of tumour progression, development of scar tissue and secondary spread of tumour cells.<sup>8</sup>

Despite the extensive evidence present to support surgery, the proximity of the tumour to vital structures in this patient called for radiation therapy and chemotherapy as the initial treatment. However, due to poor tumour regression and persistence of clinical signs, the patient eventually had to be operated. The resection was otherwise technically successful, but meningitis secondary to infection spread from the surgical wound caused the patient's death a few days postoperatively. Wound dehiscence affected the superficial as well as the deep tissues of the surgical incision and was attributed to the multiple cycles of pre-operative chemotherapy and radiotherapy. The early mortality of our case supports the fact that surgery, as the initial intervention is a better option compared to radiotherapy or chemotherapy even for lesions in the brainstem close to vital structures. An alternative can be high dose proton radiotherapy tumour control and survival.<sup>8</sup> Radiosurgery has also been tried in a few cases and favourable outcomes have been reported with local tumour control rates of up to 80% at 5 years of follow-up.<sup>10</sup> These treatment modalities may have yielded a better outcome but based on the resources available we managed the patient as described above.

There are few non-traumatic causes of intracranial haemorrhage in young patients. These include arteriovenous malformations, hypertension, ruptured aneurysms, and sympathomimetic drugs. Even though arteriovenous malformations are the most common

cause, they account for less than one third of haemorrhages in young adults. Therefore, other causes should be sought in young patients. Pontine haemorrhage, in young adults, has been seen to be a consequence of pheochromocytoma and chordoma.<sup>1,3,7</sup> This case report shows that intracranial chondrosarcoma should also be included in the differential diagnosis of pontine haemorrhage in young adults.

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