CASE REPORT

Tumour bleed manifesting as spontaneous extradural haematoma in posterior fossa

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SUMMARY

We report a unique case of primary extradural angiosarcoma of posterior fossa manifesting as extradural haematoma in a 12-year-old boy who presented with acute onset headache, vomiting, nuchal rigidity and altered sensorium. The patient underwent a retromastoid suboccipital craniotomy on emergency basis, and the lesion was excised completely. Histopathology and immunohistochemistry revealed an angiosarcoma, following which radiation therapy was given. The patient showed complete clinical and neurological improvement. At a follow-up of 2 years he is in good health without any sign of regrowth.

BACKGROUND

Angiosarcoma is a tumour of vascular origin with occurrence at various sites in the body. Primary intracranial angiosarcomas are exceedingly rare. Only 20 cases have been reported so far.¹ Extradural location of angiosarcoma presenting with apoplexy has never been reported. Our case is the first primary malignant intracranial tumour causing extradural bleed and first case of tumour causing extradural bleed in the posterior fossa.

CASE PRESENTATION

A 12-year-old boy was brought to the emergency department in altered sensorium with a history of abrupt onset headache since 3 days and vomiting for the past 2 days. There was no history of trauma or fever. On neurological examination, he had bradycardia and nuchal rigidity.

INVESTIGATIONS

A plain CT of the brain revealed an extradural hyperdense mass lateral to left cerebellar hemisphere (figure 1A). On postcontrast imaging the lesion was non-enhancing (figure 1B, C). There was mass effect in the form of oedema of underlying cerebellar hemisphere along with mild compression of the fourth ventricle. MRI of the brain showed the mass to be completely extradural. It was predominantly isointense on T1-weighted, heterogeneously hyperintense on T2-weighted image with peripheral hypointense rim of dura surrounding the lesion. A focal T2 hypointense area was seen within it which showed blooming on gradient echo images suggestive of haemorrhage (figure 1D–H). Haematological parameters were normal.

TREATMENT

In view of features of raised intracranial pressure, the patient was subjected to excision of lesion on

emergency basis by left retromastoid suboccipital craniotomy. There was an acute extradural haematoma surrounded by thinned out fleshy tissue without infiltration of bone or dura. Complete excision was performed.

OUTCOME AND FOLLOW-UP

The patient had a dramatic recovery after the surgery. Histopathological examination of the specimen revealed a lesion composed of irregular vascular spaces lined by pleomorphic endothelial cells (figure 2A–C). Numerous mitoses were present. Immunohistochemistry showed cytoplasmic membrane immunoreactivity for CD31 (figure 2D). These findings established a conclusive diagnosis of angiosarcoma. On further clinical and imaging workup (whole body positron emission tomography scan, CT of the chest and abdomen), no other focus of angiosarcoma was detected following which radiotherapy was given. On a follow-up of 2 years, the patient is in good health with no signs of recurrence (figure 1I).

DISCUSSION

Spontaneous (non-traumatic) extradural haemorrhage caused by neoplasm is rare, only 26 cases including the present case have been reported (table 1). Out of 26 cases 15 were metastatic tumours most commonly from hepatocellular carcinoma.^{2–5} In the rest 11 cases of primary intracranial tumours, 9 were of eosinophilic granuloma and 2 were of intradiploic epidermoids.^{5–8} So our case is the first primary malignant intracranial tumour causing extradural bleed. Nine patients were paediatric. Only one case of eosinophilic granuloma causing occipital extradural bleed extending to posterior fossa has been described otherwise all others had caused supratentorial bleed (table 1). Our case is the first paediatric case of tumour manifesting with isolated posterior fossa extradural haematoma.

Primary intracranial sarcomas are uncommon and comprise only 1–2% of primary intracranial neoplasms. Of these tumours, angiosarcoma is particularly rare, accounting for less than 1% of all sarcomas.⁹ These can present in the form of pure neoplasm without any other component, in association with glial elements as in gliosarcoma or as metastasis.¹⁰ The most common location of primary intracranial angiosarcoma is in the parietal lobe. Associations with chronic lymphoedema, local trauma, previous irradiation, vascular malformations, exposure to vinyl chloride, chemotherapy, arsenic exposure and the diagnostic use of

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Figure 1 (A) Plain axial CT of the brain showing an extra-axial well-defined rounded hyperdense lesion along the lateral surface of left cerebellar hemisphere with moderate perilesional oedema. There is effacement of basal cisterns and compression on the fourth ventricle. Postcontrast axial (B) and coronal (C) CT images showing no enhancement. Axial T2-weighted image (WI) (D), coronal fluid-attenuated inversion recovery (E) and sagittal T2-WI (F) showing a T2-heterogeneously hyperintense mass with a hypointense peripheral rim of dura. The lesion blooms on gradient echo image (G) while it is isointense on T1-WI. Postoperative axial T2-WI shows no residual tumour (I).

thorium dioxide as a contrast dye for angiography have been reported.¹

The onset of symptoms was abrupt in our case due to haemorrhage with aggressive clinical course. On imaging these appear as well demarcated lesions with haemorrhagic characteristics.¹ However the non-specific imaging appearances render preoperative diagnosis of primary intracranial angiosarcoma extremely difficult. The diagnosis relies on histopathological and immunohistochemical studies. On histopathology, angiosarcoma has irregular anastomosing vascular spaces lined by malignant endothelial cells. Moreover, cellular atypia, as well as many mitoses and necrosis, are histological characteristics often attributed to angiosarcomas. Immunohistochemical studies, including assays for factor CD31, CD34, factor-related antigen and

Ulex europaeus, are necessary to identify the endothelial nature of the neoplastic cells.^{1 9 11} These are useful for differentiating angiosarcoma from meningeal hemangiopericytoma, anaplastic gliomas, metastatic carcinoma, amelanotic melanoma, hemangioblastoma and several other highly vascular sarcomas.⁹

The outcome of primary cerebral angiosarcoma is generally poor. A median survival rate of only 6–10 months has been reported.^{9 11} Spontaneous intratumoural bleed is a well-described entity. Intratumoural haemorrhage has been postulated to be due to rapid tumour cell proliferation, abundant neovascularisation, and necrosis of blood vessels.^{12 13} In our case, we believe that the fragile, dysplastic thin-walled vessels of the tumour tissue along with tumour necrosis would have led to bleeding.

Figure 2 Photomicrographs show large pleomorphic tumour cells intricately admixed with blood-filled spaces and inflammatory cell infiltrate (A—H&E $\times 200$; B—H&E $\times 400$). The tumour showing frequent mitoses (as shown by the arrows in C—H&E $\times 400$) and cytoplasmic membrane immunoreactivity for CD31 (D—immunohistochemistry $\times 400$).

Table 1 Tumours presenting with extradural bleed

Author	Age/sex	Nature of the tumour	Site of EDH
Abu-Samra <i>et al</i> (1980) ⁶	11/F	Frontal intradiploic epidermoid	Frontal
Anegawa <i>et al</i> (1989) ⁵	32/F	Parieto-occipital dural metastasis of undifferentiated ovarian carcinoma	Parieto-occipital
Sato <i>et al</i> (1991) ⁵	62/M	Left frontal bone metastasis of adenocarcinoma lung	Frontal
Nakagawa <i>et al</i> (1992) ²	52/M	Occipital metastasis of HCC	Occipital
Nakao <i>et al</i> (1992) ²	58/M	Frontal bone metastasis of HCC	Frontal
Lee <i>et al</i> (1996) ²	65/F	Skull metastasis from HCC	—
Yamashita <i>et al</i> (1997) ⁵	19/F	Occipital bone Ewing's sarcoma	Occipital
Simmons <i>et al</i> (1999) ⁵	67/M	Parietotemporal dura, muscle and skin metastasis of small cell carcinoma lung	Parietal
Endo <i>et al</i> (1999) ²	44/F	Frontoparietal epidural metastasis of HCC	Combined subdural and extradural parietal bleed
Leung <i>et al</i> (1999) ⁵	—	Radiation-induced malignant fibrous histiocytoma	—
Lee <i>et al</i> ⁷	8/M	Occipital eosinophilic granuloma	Occipital
Hayashi <i>et al</i> (2000) ²	70/M	Parietal bone metastasis of HCC	Parietal
Cho <i>et al</i> (2001) ⁷	2/M	Occipital eosinophilic granuloma	Occipital
McIver <i>et al</i> (2001) ²	50/M	Parietal bone metastasis of HCC	Parietal
Chen <i>et al</i> (2002) ⁵	2/M	Occipital eosinophilic granuloma	Occipital
Martinez-Lage <i>et al</i> (2002) ⁷	9/M	Occipital eosinophilic granuloma	Occipital
Mut <i>et al</i> (2004) ⁷	9/M	Occipital eosinophilic granuloma	Occipital extending to posterior fossa
Shamim <i>et al</i> (2005) ⁵	70/M	Parietal dural metastasis of adenocarcinoma lung	Parietal
Wani <i>et al</i> ⁶	60/M	Parieto-occipital intradiploic epidermoid	Parieto-occipital
Kanai <i>et al</i> ³	—	Parieto-occipital bone metastasis of HCC	Parieto-occipital
Hassan <i>et al</i> ⁵	55/F	Parietal dural metastasis of squamous cell carcinoma lung	Parietal
Lee <i>et al</i> ⁷	10/M	Midline frontal eosinophilic granuloma	Midline frontal
Woo <i>et al</i> ⁴	46/M	Temporal skull base metastasis of HCC	Temporoparietal
Bhat <i>et al</i> ⁸	10/M	Parietal eosinophilic granuloma	Parietal
Kim <i>et al</i> ²	53/M	Temporal skull base metastasis of HCC	Temporal
Present case	12/M	Posterior fossa extradural angiosarcoma	Posterior fossa

EDH, extradural haematoma; F, female; HCC, hepatocellular carcinoma; M, male.

Learning points

- ▶ Primary intracranial angiosarcoma may rarely present with apoplexy in extradural location.
- ▶ In cases of spontaneous extradural haematoma careful scrutiny of the specimen for identification of underlying malignancy is essential.
- ▶ This is necessary for comprehensive postoperative care in the form of adjuvant radiotherapy and chemotherapy and for search of other foci of malignancy to prevent recurrence.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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