

Primary Cerebral Rhabdomyosarcoma Presenting as Haemorrhagic Stroke

Intrazerebrale Blutung als Erstmanifestation eines primären zerebralen Rhabdomyosarkoms

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Key words

- intracerebral haemorrhage
- rhabdomyosarcoma
- intracerebral tumour

Schlüsselwörter

- intrazerebrale Blutung
- Rhabdomyosarkom
- Hirntumor

Bibliography

DOI 10.1055/s-2007-1004581

Published ahead of print:

29 April 2008

Zentralbl Neurochir 2008;

69: 93–95

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Stuttgart · New York

ISSN 0044-4251

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Abstract

Intracerebral haemorrhage (ICH) occurs mostly in the context of arterial hypertension, with typical localisations. Tumour-associated bleeding is the cause of 6–10% of ICHs, mostly from metastases. We present the case of a 40-year-old female admitted originally for neck pain of sudden onset, accompanied by nausea and marked right arm paresis. A CT-scan revealed left fronto-central cortico-subcortical haemorrhage. Cerebral angiography was normal. Two months after the initial event the residual paresis worsened and the patient developed neuropsychological deficits. A CT-scan showed oedema around the original bleeding site, on MRI a solid lesion with a diameter of 5 cm could be seen, with some cystic alterations and contact to the meninges. The tumour was surgically removed, and removal at the time was considered complete. Histological analysis proved it to be an embryonal rhabdomyosarcoma. The patient's neurological deficits gradually improved. Almost three months after the operation she complained of intense left-sided headache. On CT a hyperdense left fronto-central lesion with positive enhancement could be seen; MRI confirmed a relapse tumour and showed bleeding in the rostral portion of the tumour as well as oedema. The patient started radiation therapy with a total dose of 60 Gy. Whole body image studies at the time failed to reveal any other neoplastic lesions. Two months later a CT-scan showed continued tumour growth. We present this case as a rare aetiology of intracerebral haemorrhage, more frequently associated with arterial hypertension or vascular pathology, as well as being an unusual manifestation of embryonal rhabdomyosarcoma, rarely found in the brain. The case also serves to illustrate the importance of a thorough diagnosis including MRI imaging in patients with so-called atypical ICH.

Zusammenfassung

Intrazerebrale Blutungen (ICB) treten meist im Zusammenhang mit arteriellem Hypertonus an typischen Stellen auf. Tumoren, insbesondere Metastasen, liegen 6–10% aller ICB zugrunde. Wir stellen eine 40jährige Frau vor, die sich wegen akuter Nackenschmerzen mit Übelkeit und Parese des rechten Armes vorstellte. Im CT kam eine links fronto-zentrale, kortiko-subkortikale Blutung zur Darstellung. Die zerebrale Angiografie erbrachte keine Anomalien. Zwei Monate nach dem primären Ereignis verschlechterte sich die residuale Parese erneut und die Patientin entwickelte neuropsychologische Defizite. Im CT zeigte sich ein Ödem um die initiale Blutungsstelle, in der Kernspintomographie (MRI) eine solide Läsion von 5 cm Durchmesser mit zystischen Anteilen und Kontakt zu den Meningen, die als Tumor, am ehesten Oligodendrogliom eingestuft wurde. Der Tumor wurde makroskopisch komplett entfernt. Histologisch wurde ein embryonales Rhabdomyosarkom diagnostiziert. Die Patientin erholte sich gut. Fast drei Monate später klagte sie über starken linksseitigen Kopfschmerz. Im CT kam eine hyperdense, Kontrastmittel aufnehmende Läsion links fronto-zentral zur Darstellung, in der MRI wurde das Tumorrezidiv mit Einblutung in seinen rostralen Anteilen bestätigt. Es wurden keine neoplastischen Läsionen in anderen Körperpartien gefunden. Die Patientin begann eine Strahlentherapie mit einer Gesamtdosis von 60 Gy. Zwei Monate später kam in der Kontrollbildgebung fortgesetztes Tumorwachstum zur Darstellung. Wir stellen den Fall vor, da er einerseits eine seltene Ätiologie einer ICB darstellt, andererseits eine höchst außergewöhnliche Manifestationsform eines embryonalen Rhabdomyosarkoms, welches selten im Gehirn auftritt. Der Fall dient auch dazu, die Bedeutung einer erschöpfenden Bildgebungsdiagnostik inklusive MRI bei allen Patienten mit sogenannt atypischen ICB zu unterstreichen.

Introduction

Rhabdomyosarcoma (RMS) is a rare tumour occurring predominantly in the paediatric age group. Cerebral rhabdomyosarcomas are even rarer, presenting usually as slowly progressive neurological deficits [4,6]. Intracerebral haemorrhage (ICH) occurs mostly in the context of arterial hypertension, with typical localisations. Tumour-associated bleeding is the cause of 6–10% of ICHs [8].

Case report

A 40-year-old female presented with sudden neck pain, nausea, and marked right arm paresis. A CT-scan (◉ Fig. 1) revealed left fronto-central cortical-subcortical bleeding. Angiography did not show any arterio-venous malformations or aneurysms. The only previous disease known was dietetically treated diabetes. She improved with conservative treatment.

Two months later the patient presented with right hemiparesis, aphasia and mild neuropsychological impairment. A CT-scan showed a mixed hypo- and hyperdense lesion and oedema around the original bleeding site. MRI (◉ Fig. 2a, b) showed a solid lesion with some cystic alterations and broad contact to the meninges, considered a probable oligodendroglioma. Neurosurgical intervention resulted in macroscopically total tumour resection. Histologically a small cell tumour with scattered rhabdomyoblasts was found. Immunohistochemical

staining was positive for muscle actin and desmin, resulting in a diagnosis of embryonal rhabdomyosarcoma (◉ Fig. 3a, b). Adjunctive chemo- and radiotherapy was advised, but rejected by the patient's husband.

After three months the patient still had severe organic brain syndrome, aphasia and spastic right hemiparesis. Control CT showed a left fronto-central hypodense lesion with slim contrast enhancement close to the skull (◉ Fig. 4a), suggesting the presence of residual tumour tissue.

Another month later the patient communicated increasing left headache. On CT the lesion was larger and had become hyperdense (◉ Fig. 4b). MRI confirmed a relapse tumour with haemorrhage. Spiral CT scans of the thorax, abdomen and pelvis failed to reveal any other neoplastic lesions. The patient's husband (and legal guardian) continued to refuse chemotherapy, but accepted radiotherapy, so she was treated with a total dose of 60Gy.

After 11 months of clinical stability and only very slight tumour growth on MRI, symptoms worsened. MRI then revealed marked tumour growth. Treatment was restricted to palliative measures.

Discussion

ICH typically is caused by the rupture of small perforating vessels in the context of arterial hypertension in 45–65% of cases

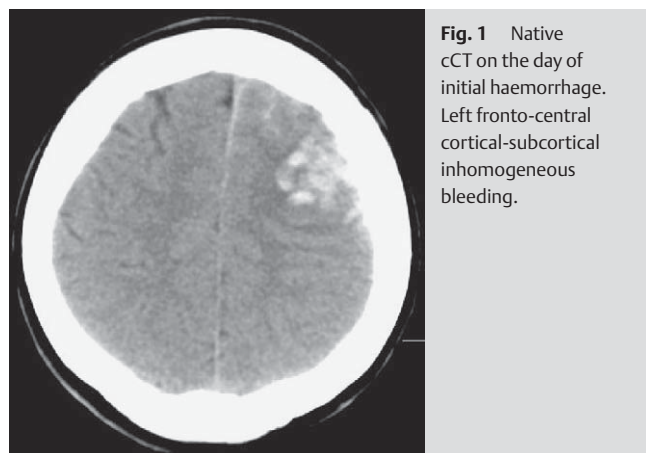


Fig. 1 Native cCT on the day of initial haemorrhage. Left fronto-central cortical-subcortical inhomogeneous bleeding.

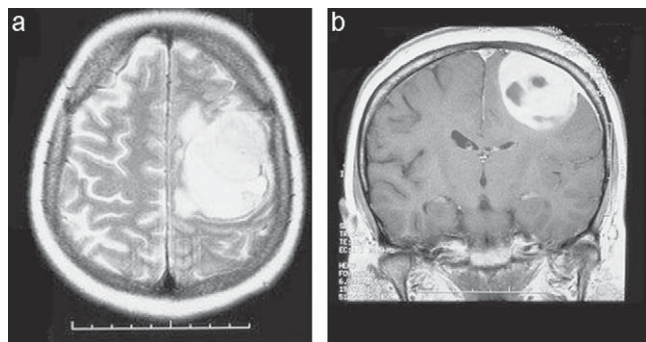


Fig. 2 MRI two months after initial haemorrhage: **a** axial plane T2-FSE, and **b** coronal plane T1-SE contrast-enhanced (15 ml Magnevist) MRI demonstrating high left fronto-central solid lesion, with a diameter of 5 cm and cystic alterations, broad contact to the meninges, and a small surrounding oedema.

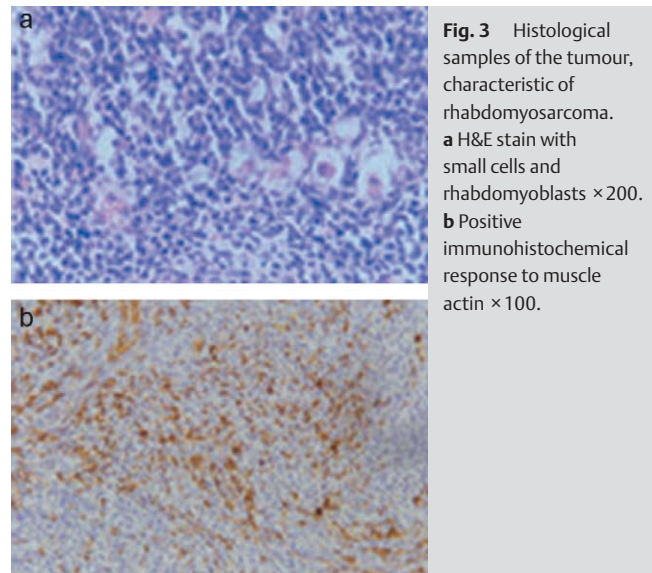


Fig. 3 Histological samples of the tumour, characteristic of rhabdomyosarcoma. **a** H&E stain with small cells and rhabdomyoblasts $\times 200$. **b** Positive immunohistochemical response to muscle actin $\times 100$.

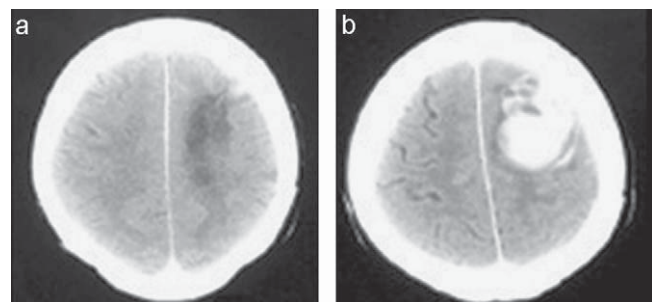


Fig. 4 **a** Native cCT five months after initial haemorrhage: hypodense lesion with meningeal and parameningeal enhancement after administration of contrast medium, indicative of residual tumour. **b** Native cCT six months after initial haemorrhage: tumour growth and recurrent haemorrhage.

[3, 8]. The localisation of ICH is one third lobular, one third in the putamen, 12–26% in the thalamus, 5% in the caudate nucleus, 10% cerebellar and 8–10% pontine [7].

Among the non-hypertensive causes are amyloid angiopathy, vascular malformations, aneurysms, tumours, anticoagulant agents, systemic bleeding disorders, septic embolism, vasculitis and sympathomimetic drugs [8].

Lobular ICH is related mostly to cerebral amyloid angiopathy, tumours or sympathomimetic drugs.

Tumour bleeding occurs in 3.5% of all patients with intracerebral tumours (2.4% of primary brain tumours and 9.3% of brain metastases, mainly from thyroid papillary carcinomas, hepatocellular carcinoma, melanomas, hypernephroma, choriocarcinoma and bronchogenic cancer) [3, 8, 9].

Rhabdomyosarcoma (RMS) is a tumour of mesenchymal origin, which represents 13% of all paediatric malignancies (incidence: 4 per million in patients aged < 15 years) [4, 5]. Primary intracranial rhabdomyosarcoma is extremely rare and occurs almost exclusively in the paediatric age group [5, 6]. On immunohistochemical evaluation RMS shows a positive reaction for muscle actin, myoglobin and desmin [6, 13].

The prognosis of localised rhabdomyosarcoma is good with adequate treatment, but poor when – like in our patient – the meninges are involved, due to poor response to systemic chemotherapy [14]. Metastatic involvement of the brain implies a mean survival time of 2.7 months [12]. Radiation can improve the prognosis of these patients [14]. In the present case, the husband's refusal of adjunctive treatment, particularly radiotherapy, may have contributed to the unfavourable outcome.

To our knowledge, primary intracranial rhabdomyosarcoma had been reported only five times so far [2, 4–6, 11]. ICH as a first manifestation has been described only once, in a metastatic lesion [1]. To our knowledge, this is the first published case of ICH as an initial manifestation of primary cerebral RMS.

Native or contrast-enhanced CT is always diagnostic of ICH, but may miss underlying causes such as neoplastic lesions. MRI has a higher resolution and is capable of showing the dynamic evolution of the haematoma as well as the nature of underlying lesions [3, 8, 10]. Depending on the localisation and evolution of the haemorrhage, MRI should be performed to achieve a differential diagnosis of the underlying causes as soon as possible. In a very large haemorrhage early MRI may miss an underlying tumour, therefore in these cases MRI should be repeated after approximately one month or be postponed until that time.

In the present case the patient suffered a high left frontal-central haemorrhage, by its localisation very suspicious for an underlying neoplastic lesion. As it was not very large, early MRI might very well have identified the cause at that point in time.

Conclusions



In this patient a more thorough initial investigation including MRI might have led to an earlier diagnosis and adequate neurosurgical and radiation therapy, which could have resulted in a better prognosis.

Any patient bleeding in an atypical localisation should be submitted to complete neuroradiological diagnostic procedures including contrast-enhanced CT, angiography and MRI for further differential diagnosis.

Conflict of interest: None

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