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CASE REPORT

Sudden death due to subarachnoid bleeding from ecchordosis physaliphora

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Abstract Ecchordosis physaliphora (EP) is a rare intracranial mass derived from ectopic notochordal tissue. It is usually a fortuitous finding at autopsy or by computed tomography/magnetic resonance imaging. Very few authors have described an EP-associated symptomatology. In this study, we report a case of the sudden and unexpected death of a 48-year-old woman. At autopsy, the cause of death was subarachnoid bleeding, the origin of which was identified as a gelatinous mass stemming from the dura mater and occupying the prepontine space. Further histological and immunohistochemical investigations allowed the diagnosis of EP.

Keywords Sudden death · Subarachnoid bleeding · Ecchordosis physaliphora

Introduction

Ecchordosis physaliphora (EP) is considered to be the remains of the notochord and consists of a small gelatinous mass, typically located intradurally in the prepontine cistern, attached to the dorsal wall of the clivus [7, 14]. The frequency of this finding varies between 0.5 and 2% of autopsies [8]. EP is usually asymptomatic and only few authors have reported EP-associated symptoms due to tumour expansion and compression of the surrounding structures [1, 12, 13]. Fatal subarachnoid bleeding due to rupture of an EP has been described only once [14].

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Case report

A 48-year-old woman suddenly complained of splitting headache during a cycle tour and collapsed while dismounting from the bicycle. The emergency physician could only certify death. The husband reported a completely negative past history.

On external examination, the body was that of a middle-aged Caucasian woman, 163 cm in height and 65 kg in weight. Abrasions due to the collapse were detected at the right knee and elbow.

The internal examination of the head showed a moderate subarachnoid bleeding over the hemispheres and the basis. A 3.2×2.2×0.4 cm in size, white-greyish, soft lobular gelatinous tissue originating from the *dura mater* occupied the space above the *sella turcica*, behind the pituitary stalk (Fig. 1). This tumour did not infiltrate the surrounding vessels or the nervous parenchyma. The circle of Willis was carefully investigated and did not show any disorders, i.e. aneurysms or ruptures. Other autopsy findings were severe brain oedema, moderate aorto-coronary sclerosis and a mild bronchitis with some mucous.

The death was caused by nervous dysregulation due to the subarachnoid bleeding. The brain was collected in toto and fixed in 4% buffered formalin for 4 weeks before neuropathological investigation.

The histology of the tumour tissue showed cell-poor lobules. The cells had a large eosinophilic cytoplasm with intracytoplasmic mucous droplets (physaliphorous cells) (Fig. 2). Neither mitoses nor infiltration of the surrounding

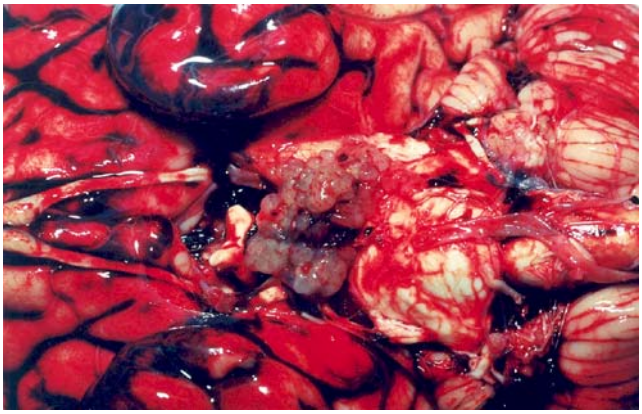

Fig. 1 Subarachnoid bleeding originating from ecchordosis physaliphora

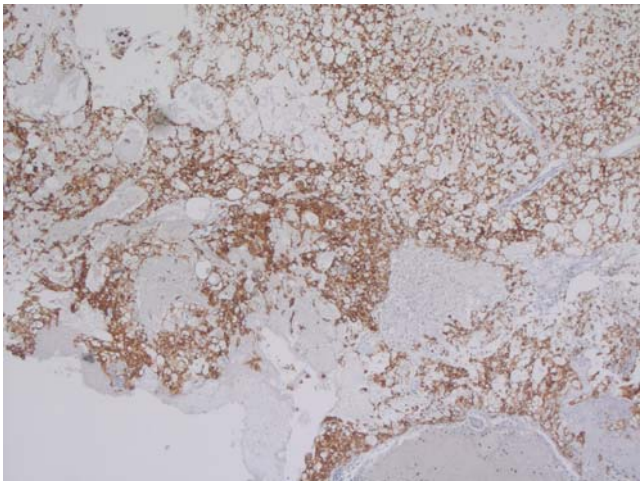

Fig. 3 The cells express the epithelial marker cytokeratin (clone KL1, ×40)

vessels or brain parenchyma were visible. By immunohistochemistry, the vast majority of the cells expressed the epithelial markers cytokeratin (clone KL1; Fig. 3) and epithelial membrane antigen. Some cells were positive for vimentin and S-100 protein. The reaction with the proliferation marker MIB-1 was negative. In addition to fresh, space-occupying bleeding within the ecchordosis (Fig. 4), haemosiderin deposits indicating older haemorrhages were also visible (Fig. 5). Serial sections of the arteries of the basal brain did not show any pathological changes.

The neuropathological investigation allowed the diagnosis of a subarachnoid bleeding originating from an EP.

Discussion

The notochord represents the primitive skeleton of the vertebrates and appears at the third gestational week and gives rise to the nucleus pulposus of the intervertebral discs at maturity. EP is a congenital malformation originating from ectopic vestiges of notochordal tissue. The typical localisation is the prepontine cistern, but it can be found all along the vertebral column.

The histology of EP is very similar to that of intradural chordoma, its malignant counterpart [5, 7, 12] and both present physaliphorous cells arranged in a myxoid matrix. Immunohistochemistry is useful to support the diagnosis [9]. In particular, the low proliferative activity supports the diagnosis of EP [2, 7, 12].

As the basal arteries are the most common origin of subarachnoid bleeding [4], they were carefully dissected, but neither the macroscopic or the histological investigations revealed any pathological changes, thus excluding this as the origin. However, fresh haemorrhages as well as haemosiderin deposits were detected within the EP, which was assumed to be the origin of the bleeding.

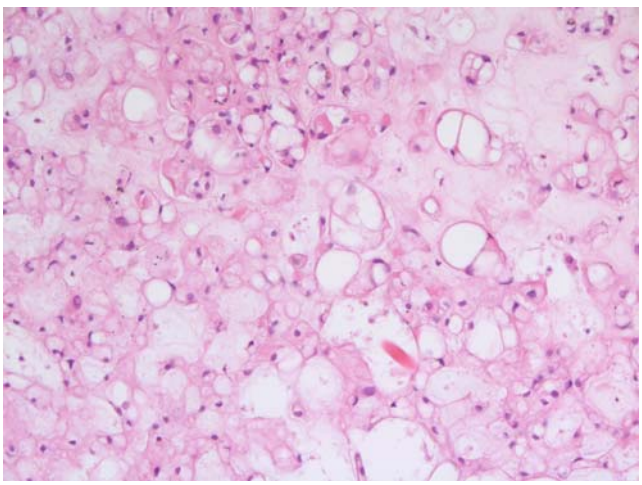

Fig. 2 Physaliphorous cells with large cytoplasma and introcytoplasmatic mucous droplets (H&E, ×200)

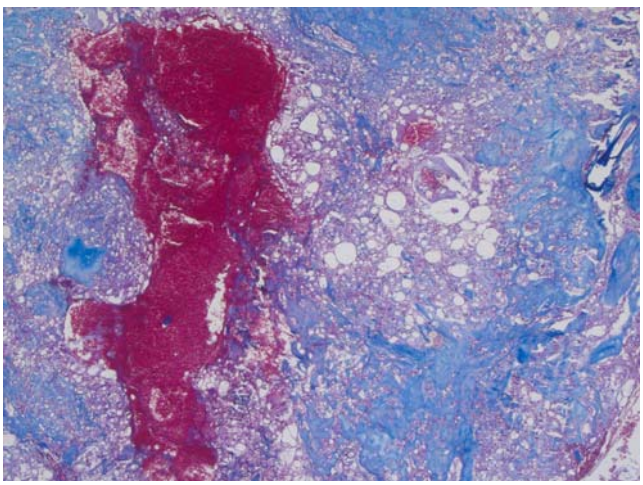

Fig. 4 Fresh, space-occupying haemorrhage within the ecchordosis (Azan, ×40)

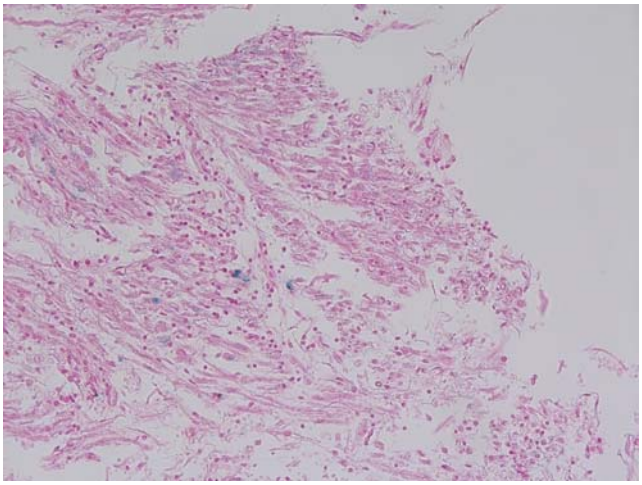

Fig. 5 Haemosiderin deposits indicating older haemorrhages within the ecchordosis (Prussian blue, ×100)

Like similar publications [3, 6, 10, 11], this manuscript aims at making the medico-legal profession aware of a rare condition that can lead to sudden death.

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