Ecchordosis physaliphora – a case report and a review of notochord-derived lesions

Ecchordosis physaliphora – opis przypadku i omówienie patologicznych pochodnych struny grzbietowej

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Abstract

Some notochord cells remain along the axis of the vertebral column after embryogenesis. These 'notochordal remnants' have some similarities, but their biological behaviour varies considerably. They can give rise to benign lesions such as ecchordosis physaliphora (EP) and 'benign notochordal cell tumour' (BNCT), or aggressive ones like chordoma.
We review the problems of the differential diagnosis of notochordal remnants apropos of a case of the incidental autopsy finding of EP in a 78-year-old man, who died due to heart infarction. The 6-mm asymptomatic gelatinous lesion was fixed to the basilar artery on its ventral aspect.
Small EPs can be easily overlooked in autopsy. Ecchordosis physaliphora and intradural chordoma share some similarities that may be misleading and may even result in the wrong diagnosis and therapy. The recently reported new entity BNCT poses a similar problem. We review the literature illustrating the most important features of notochord-derived lesions and discuss the relationships between these lesions with regard to molecular genetics.

Key words: chordoma, notochord, basilar artery, nucleus pulposus, autopsy.

Streszczenie

Podczas formowania się jąder miażdżystych na pozostałościach komórek struny grzbietowej rozwijają się zmiany podobne do siebie, mające jednak różny przebieg kliniczny. Niektóre z nich, takie jak *ecchordosis physaliphora* (EP) i *notochordal benign cell tumour* (NBCT), mają charakter łagodny, inne (np. struniak) – agresywny. Celem pracy jest przegląd najważniejszych cech zmian wywodzących się ze struny grzbietowej i ich diagnostyka różnicowa na podstawie przypadku EP stwierdzonego podczas autopsji mózgu 78-letniego mężczyzny zmarłego na atak serca. Galaretowaty guz o największym wymiarze 6 mm, przytwierdzony do brzusznej części tętnicy podstawnej nie dawał żadnych objawów.
Ecchordosis physaliphora to zazwyczaj mały, bezobjawowy guzek, łatwy do przeoczenia podczas autopsji. Struniak i EP wywodzą się z tych samych komórek, lecz ze względu na podobieństwa mogą być ze sobą mylone, co skutkuje niewłaściwym rozpoznaniem i ewentualnym leczeniem. Podobny problem występuje w przypadku NBCT. W pracy poruszono również kwestię ich wzajemnego związku na podstawie badań genetycznych.

Słowa kluczowe: struniak, struna grzbietowa, tętnica podstawna, jądro miażdżyste, autopsja.

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Introduction

The notochord develops in humans during the third week of embryonic life and persists in adults as the nucleus pulposus of the intervertebral discs. During formation of the nucleus pulposus, some cells of the notochord remain along the axis of the vertebral column. They can create clinically important lesions ranging from ecchordosis physaliphora, through benign notochordal cell tumours (BNCTs), to a rare type of tumour called chordoma. These 'notochordal derivatives' have a lot of pathological similarities, but their biological behaviour varies considerably. The problem of their differential diagnosis affects the modalities of clinical treatment.

Case report

During the autopsy of a 78-year-old man, who died due to heart infarction, a lesion 6 × 4 × 3 mm, fixed to the basal artery on its ventral site, without infiltration of the artery, was found. It was well circumscribed, glossy, with gelatinous consistency (Fig. 1).

Microscopically, the lesion was formed by loosely packed cells with bland nuclei and ample vacuolated ('physaliphorous') cytoplasm dispersed within a myxoid background (Figs. 2 and 3). Immunohistochemical staining showed positivity for S-100, neuron-specific enolase (NSE), and epithelial membrane antigen (EMA) (Fig. 4). No mitoses were visible and immune labelling for topoisomerase-2-alpha (proliferative anti-

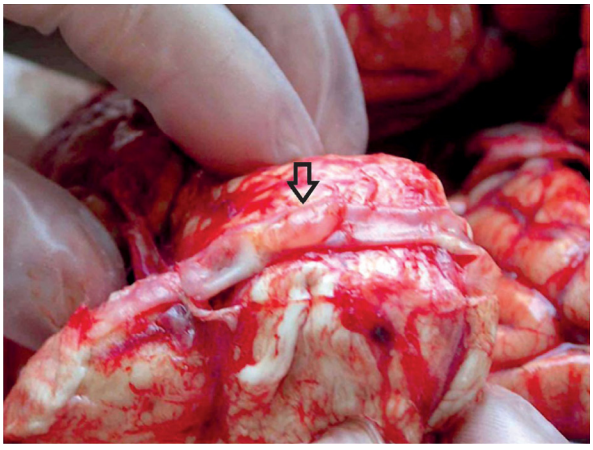

Fig. 1. Macroscopic *in situ* picture of ecchordosis physaliphora. Small jelly-like tubercle attached to basilar artery (arrow)

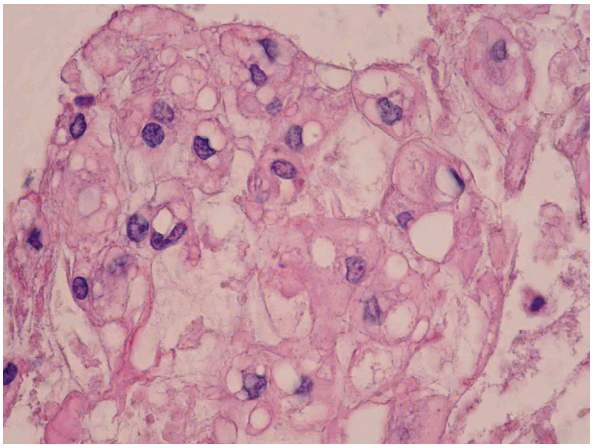

Fig. 3. High power picture of ecchordosis physaliphora. The most characteristic cells, called physaliphorous, have numerous, small intracytoplasmic vacuoles (HE)

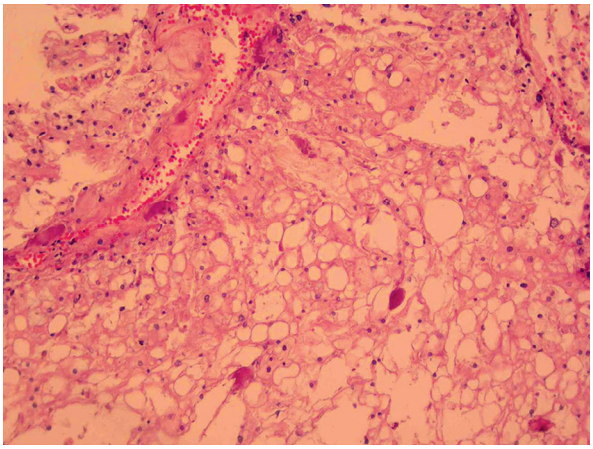

Fig. 2. Microscopic picture of ecchordosis physaliphora shows a tissue composed of cells with ample, sometimes strongly vacuolated cytoplasm without distinct borders, with small, bland nuclei, focally interspersed within myxoid matrix (haematoxylin-eosin, HE)

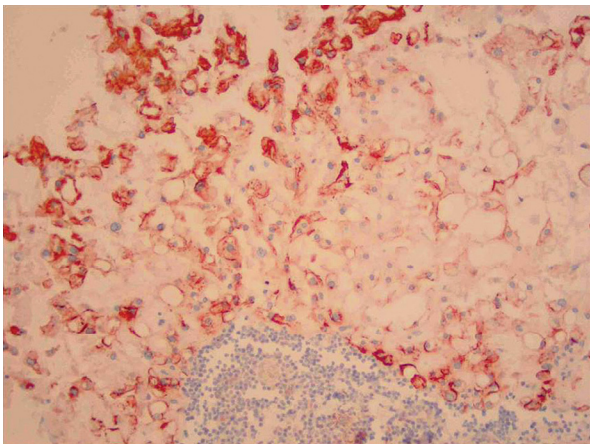

Fig. 4. Immunohistochemistry with antibody against EMA shows strong positivity. Noteworthy are the clusters of lymphocytes that can be seen in the lower part of the picture, which may suggest a form of immune response to the lesion