CASE REPORT

An Incidental Finding of Ecchordosis Physaliphora in a Case of Abducens Nerve Palsy: Case Report

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ABSTRACT

Ecchordosis physaliphora (EP) is a rare non-malignant mass that originates from remains of the notochord and is typically asymptomatic. A 42-year-old man presented with sudden onset of painless horizontal diplopia and his neurological exam showed sixth cranial nerve palsy. Magnetic resonance imaging (MRI) identified a non-enhanced retroclival mass (EP) with increased signal intensity on T2 and decreased signal intensity on T1-weighted sequences. He was treated with methylprednisolone, completely recovered in four weeks and has remained symptom free. Conservative management should be attempted before surgery in all cases since symptoms can resolve spontaneously and EP could be an incidental finding.

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Introduction

Ecchordosis physaliphora (EP) is usually a midline non-malignant gelatinous tumour that originates from the remnants of the notochord. It is normally a benign, asymptomatic, and intradural lesion with a stalk connected to the clivus. It is found in about 2% of all autopsies.¹ Symptomatic cases are extremely rare, and the majority are managed by surgical resection.²

Case presentation

History and physical exam

A 42-year-old, healthy man developed painless, progressive horizontal diplopia. On exam, the visual acuity corrected to 20/20, and he identified correctly all the colour plates in each eye; the intraocular pressure was normal with equal pupillary size and reaction to light. He had normal visual fields to confrontation bilaterally. Slit-lamp and direct ophthalmoscopy were unremarkable. He had a left abduction defect at presentation of −4 (100% deficit).

Investigation

Pre- and post-contrast magnetic resonance imaging (MRI) showed a mass lesion with decreased signal on T1-weight images (Figure 1A) and increased signal intensity on T2-weight images (Figure 1B) and it was non-enhancing with contrast (Figure 1C). The bulk of the mass was located towards the right of the midline, measured 8 × 5 mm, and appeared to be attached to the clivus. The abducens nerves did not enhance. Although we detected non-specific hyperintense signal foci in the cerebral white matter on T2-weighted sequences, their shape and location did not fit the imaging characteristics of multiple sclerosis; hence, we decided to explore this diagnosis further. Lumbar puncture revealed a normal opening pressure. Analysis of the cerebrospinal fluid was normal except for mildly elevated protein (63 mg/dl with a normal range of 12–60 mg/dl). Oligoclonal bands were negative. Serum folic acid, vitamin B12, thyroid stimulating hormone (TSH), protein electrophoresis, blood glucose, antinuclear antibody, anti-neutrophil cytoplasmic antibody, and myasthenia gravis also were normal.

Figure 1. Panel A: Axial T1 MRI. The arrow points to a hypointense preclival mass located towards the right of the midline. Panel B: Axial T2 MRI. The arrow shows a hyperintense preclival mass that lean towards the right of the midline. Panel C: Axial T1 contrast-enhanced MRI that shows a non-enhanced preclival lesion (*Ecchordosis physaliphora*).

Treatment

The initial concern was EP-induced compressive effect on the left sixth cranial nerve. The lesion was small and located towards the right of the clivus, opposite to the affected left abducens nerve; however, aware of the fact that MRI does not provide a precise tridimensional image of the entire clivus, we felt that compression of the left sixth cranial nerve was a still a possibility and decided to treat with methylprednisolone. Accordingly, a three-day course of intravenous methylprednisolone 1 g/day led to mild improvement of diplopia by day two. Two weeks later, he still had diplopia in primary and left gaze, though exam showed lessening of the ophthalmoplegia. The abduction deficit decreased to -3 (75% deficit). Alternate cover testing at distance showed an esophoria of right

gaze -4 PD, primary gaze -20 PD, and left gaze -35 PD. He completely recovered one month later, with orthophoria on cross-cover test.

Discussion

The first pathologic case of EP was described by Luschka in 1856, and Rippert in 1894 introduced the name “ecchordosis physaliphora.”¹ EP is a benign lesion, in contrast to the malignant clivus tumour known as chordomas. Whereas the management of asymptomatic EP is mainly conservative,³ the management of other clivus lesions, in particular chordomas, may require different combinations of surgery, chemotherapy, and radiotherapy.⁴ The histologic examination of the lesion shows physaliphorous cell nests and a pedicle composed of mature cartilaginous cells.⁵

EP tumours are slow-growing, small, mostly intradural, and rarely contrast enhancing; they may have a bony stalk and rarely cause cranial neuropathies.⁶ MRI with administration of contrast is the key diagnostic tool in differentiating EP from malignant chordomas; thus, MRI tests must include contrast administration. Both tumours show decreased signal intensity on T1-weighted MRI and increased signal on T2-weighted MRI; EP, however, does not show enhancement with gadolinium-contrast medium.⁷ The different surgical approaches for symptomatic EP depend on tumour size and location, and the aim of the operation is total resection.²

The annual incidence of abducens paralysis is 11.3/100,000.⁸ The most common causes are microvascular infarction (28%), trauma (5%), neoplasm (5%), aneurysm (4%), and others (20%), but in many instances, they are idiopathic (24%).⁹ In this age group, the possible ischaemia of the nerve core secondary to microvascular occlusion is considered a common mechanism. Patel et al. reviewed 76 cases of isolated abducens palsy and found that 51.3% of the patients had hypertension, 23.7% had diabetes, and 18.4% had diabetes and hypertension.¹⁰

Our patient had no previous medical history or risk factors for an ischaemic cranial nerve palsy and presented with painless diplopia. An isolated sixth cranial nerve palsy was diagnosed and felt to be due to a compressive lesion. MRI was able to exclude neoplastic, granulomatous, and infectious lesions, which were considerations in the differential diagnosis. Nodular skull base lesions may occur with meningeal inflammation, infectious meningitis, and lymphoma. The rapid development of unilateral abducens palsy in an otherwise healthy young man prompted us to perform lumbar puncture for cerebrospinal fluid (CSF) analysis and to measure the intracranial pressure. Multiple sclerosis is also a consideration in this age group, and isolated fascicular abducens paresis may be the first manifestation of this illness¹¹; however, we concluded that the periventricular white matter changes in this case were non-specific.

In retrospect, when considering the rightward lesion location and the speed of recovery, it is unlikely that this patient's ophthalmoplegia was caused by compression. In addition, if compression was the cause, one might expect recurrence after steroids

were stopped even though abducens neuropathies due to compression by extra-axial lesions may improve spontaneously.¹² Ahn et al. reported a case of EP with diplopia where patient responded to oral steroids, with recurrent diplopia, six months later.¹³ Our young patient has been symptom free for more than one year. As it stands, we are not able to categorically determine the cause of the ophthalmoplegia in this case, and idiopathic sixth cranial nerve palsies typically have a high rate of recovery. In the absence of a known aetiology, overall prognosis for monophasic idiopathic abducens palsy is very good. King et al. reported about 78% spontaneous recovery with a mean recovery time of 9.7 weeks.¹⁴ Our patient spontaneously recovered completely within 4 weeks and remains symptom free for more than 1 year. He will continue annual clinical and imaging surveillance monitoring. In addition to abducens compression, mass effect on the basilar artery may be an additional reason for intervention.⁵

In conclusion, EP is mainly an asymptomatic lesion; its presence in a patient with a sixth nerve palsy is not necessary a causation. MRI characteristics separate it from chordomas and other malignant clivus tumours. Initially, conservative management should be attempted, unless compelling evidence of lack of improvement or worsening develops. The role of steroids in the management of EP is uncertain at this time.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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