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Extra-axial Subarachnoid Ependymoma Mimicking a PCA Schwannoma

Ponto-serebeller Köşe Tümörünü Taklit Eden Ekstra-aksiyal Subaraknoid Ependimom

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ABSTRACT

The presentation, diagnosis, and treatment are described for a patient with an ependymoma involving the left cerebellopontine angle (PCA). A 34-year-old woman had left facial paralysis, hearing loss and headache in 8 months. Computerized tomography (CT) revealed a mass lesion of the left PCA which was enhanced homogeneously. Magnetic resonance imaging also (MRI) showed the mass in the left PCA which was high-intense on T1-weighted images, iso- and high-intense on T2-weighted images and homogeneously enhanced by administration of Gd-DTPA. The patient was operated on and the tumor was totally removed. Histopathological examination of the specimen revealed a typical ependymoma. The postoperative course was uneventful with complete resolution of the symptoms on admission. We present here an unusual case of PCA ependymoma located within the PCA and exhibiting no continuity with the ventricular systems. The radiological and clinical features of the tumour mimicked a PCA schwannoma.

KEY WORDS: Ependymoma, schwannoma, cerebellopontine angle

ÖZ

Sol pontoserebellar köşede ependimomu olan bir hastanın tanısı ve tedavisi sunuldu. Hasta 34 yaşında kadın, sol tarafında fasiyal paralizi, işitme kaybı ve 8 aydır başağrısı şikayeti mevcut. Bilgisayarlı Tomografisinde sol pontoserebellar köşede homojen kitle lezyonu mevcuttu. MRG' de sol pontoserebellar köşede T1 ağırlıklı kesitlerde hiperintens ve T2 ağırlıklı kesitlerde izo veya hiperintens, Gd-DTPA verildikten sonra homojen tutulum gösteren kitle lezyonu görüldü. Hasta opere edilerek tümör total olarak çıkartıldı ve histopotolojik tanı ependimom ile uyumlu olarak değerlendirildi. Postoperatif takiplerinde hastanın semptomları düzeldi. Biz bu çalışmada, radyolojik olarak schwannomu taklit eden ventriküler sistem ile ilişkisi olmayan pontoserebellar köşe lokalizasyonlu ependimom vakasını sunduk.

ANAHTAR SÖZCÜKLER: Ependimom, schwannoma, serebellopontin açı

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INTRODUCTION

Ependymomas are rare tumors arising from the cells lining the ventricular systems and the central canal of the spinal cord. They account for 1.2-6% of primary intracranial neoplasms. Approximately two-thirds are cerebral ependymomas are most of them arise within the fourth ventricle. Ependymomas can present as entirely extra-axial intracranial masses and rarely the confines of the PCA. Ependymoma has a number of variants (such as cellular, papillary and myxopapillary), but the clear cell variant is rare. The differential diagnosis therefore includes oligodendroglioma, neurocytoma and medullobastoma (2,4,8,12,) Acoustic neuromas (AN) are benign tumors and are also known as vestibular schwannomas. Their origin is the Schwann cell, comprising the myelin sheath of the vestibulocochlear nerve. Commonly, AN develop at the vestibular portion of the nerve. They can be localized within the internal acoustic meatus or the cerebellopontine angle, but they can also have both intracanalicular and cerebellopontine angle components. Commonly, the neurologic symptoms at presentation include sensorineural hearing loss, unsteadiness, dizziness, tinnitus, mastoid pain or otalgia, headache, and facial numbness. Further symptoms, including facial weakness, dysarthria, dysphagia, and hydrocephalus, are associated with larger tumors (11, 10, 14).

CASE REPORT

A 34-year-old woman was admitted in June 2003. She had left facial paralysis, hearing loss and headache for the last 8 months. Neurological examination on admission revealed only left facial paralysis. Computerized tomography (CT) revealed a mass lesion of the left PCA which was enhanced homogeneously. Magnetic resonance imaging also (MRI) showed the mass in the left PCA which was iso-intense on T1-weighted images, iso- and high-intense on T2-weighted images and homogeneously enhanced by administration of Gd-DTPA. A left retrosigmoidal craniotomy was performed. A yellow-grayish tumour was located in the PCA and was sharply demarcated from the surrounding tissue. The tumor was covered with arachnoid membrane giving an impression that tumor arose in the subaracnoid space. The tumor was totally removed. The postoperative course uneventful with complete resolution of the symptoms on admission. She was discharged 6 days after the operation. Histopathological examination of the specimen revealed a typical ependymoma.

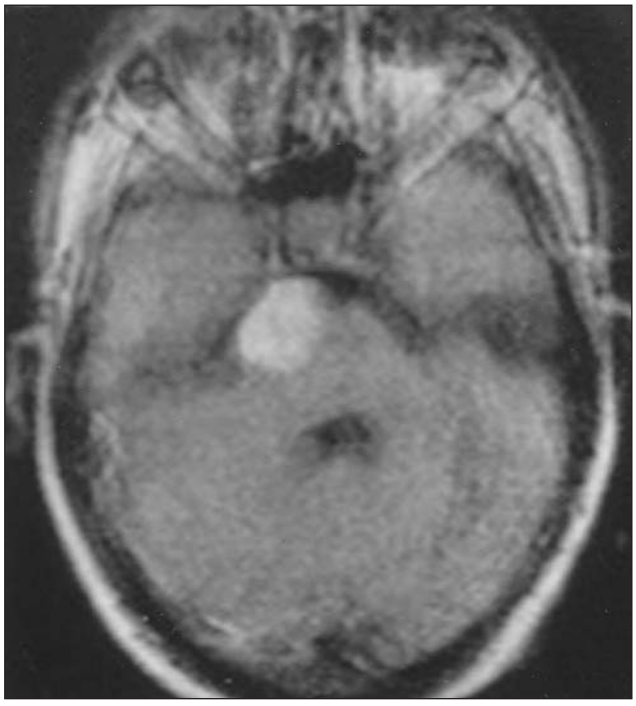

Figure 1: Preop. MRI showing the mass in the left PCA

Figure 2: Postop. MRI showing that the tumor has been totally removed.

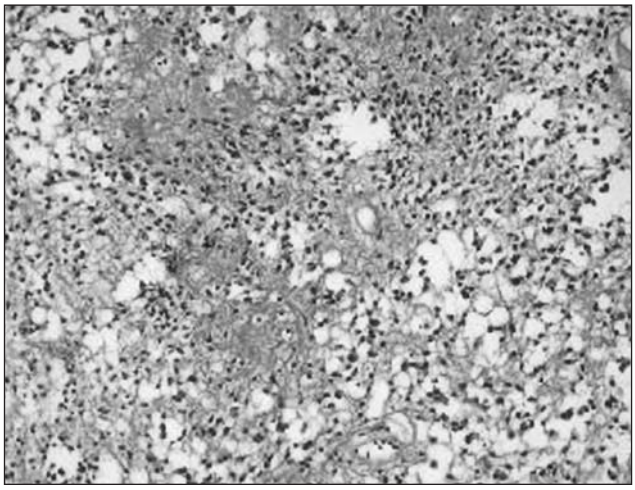

Figure 3: The cytoplasmic processes of ependymal tumor cells condense around blood vessels to form pseudorosettes (Hematoxylin and eosin stain, orginal magnification X50)

DISCUSSION

Acoustic schwannomas are benign tumors that originate from Schwann cells surrounding the vestibular nerve, usually within the internal auditory canal. (IAC) As the tumor progresses, it fills the canal and extends into the cerebellopontine angle. Acoustic schwannomas are relatively uncommon with an incidence of approximately 1 new case per 100,000. The mean age at diagnosis is approximately 45 to 47 years, and there is a slight female preponderance (10, 7).

Tumors of the cerebellopontine angle are frequent; acoustic neuromas, epidermoid tumors and meningiomas represent the great majority of such tumors. In addition, CPA lesions can be secondary to an exophytic brainstem or ventricular tumor such as ependymoma. The site of origin is the main factor in making a preoperative diagnosis for an unusual lesion of the CPA. Both acoustic neuromas and ependymomas may have similar radiologic appearances including size, contour, enhancement, and signal intensities. Tumoral extension into the IAC is a valuable diagnostic criterion for acoustic neuromas, but its not possible to detect in every patient. In such cases, making a differential diagnosis between acoustic neuroma and other CPA tumors is difficult. In our case, the lesion had regular sharp borders and showed intense homogenous enhancement which is why neuroma was thought to be the primary diagnosis although there was no IAC extension.

Ependymoma is a tumor having a relatively slow growth but with a potential for local invasion. Most intracranial ependymomas are believed to arise from the ependymal linings of the ventricular system. However, this tumor might originate from heterotopic ependymal cell rests either in the cortex or in the subarachnoid space. Aberrant ependymal cell rests can be misplaced in various parts of body during the embryonal periods(1, 3, 5). Consequently, ependymomas may occur in the ovary, mediastinum and elsewhere away from the central nervous system. Approximately 60 to 70% of ependymomas in the cranium occur within the confines of the posterior fossa, and most of these arise in the fourth ventricle (6, 13). Sato et al. envisage two possibilities regarding development. The first possibility is that the tumor has developed from an ependymal cyst. Intraparenchymal or subaracnoid ependymal cysts have been reported and thought to develop as an out-pouching from the ventricles and to be derived from ectopic ependymal nests resulting from migration disorders of the germinal matrix. The second possible pathogenic hypothesis would be to consider that the tumor would represent a primitive neuroectodermal tumor which would have differentiated extensively along the ependymal lineage (9).

To our knowledge 7 cases of extra- axial ependymoma, including 4 cases of infratentorial and 3 of supratentorial location have been reported have been previously reported (Table 1). Three cases had calcified areas and all cases had heterogeneous enhancement after contrast administration. Although radiological findings of the previously reported cases were not characteristic for meningioma and schwannoma (PCA location) all cases were preoperatively diagnosed as meningioma and schwannoma because of their location. All 7 cases evaluated by CT scan and MRI imaging had cystic degeneration.

We presented a case with an ependymoma of the left PCA. The radiological and clinical features of the tumour mimicked a PCA Schwannoma.

Table: 1 Summary of reported cases of extra-axial ependymoma

Authors	Year	Patient		Location	Radiological features						operative findings	
		Age (year)	Sex		T1W2	T2W1	Enhancement	Cyst	Calc.	Dural Tail sign	Dural attachment	Pial adhesion
Hanchey et al	1976	29	M	Interhemispheric	NA	NA	NA	NA	NA	NA	(+)	(+)
Cosgrove et al	1985	78	M	CP cistern	NA	NA	NA	(+)	NA	NA	(-)	(+)
Hayashi et al.	1994	13	M	Occipital convexity	iso	iso	heterogeneous	(+)	(+)	(-)	(-)	(+)
Fukui et al.	1996	66	M	CP cistern	low	slightly high	heterogeneous	(+)	NA	(-)	(-)	(-)
Donich et al.	1999	22	F	CP cistern and Cavernous sinus	NA	high	heterogeneous	(+)	(-)	(-)	(-)	(+)
Youkilis et al	2001	20	M	Interhemispheric	iso	iso	heterogeneous	(+)	(+)	(-)	(+)	(-)
Goto et al.	2003	29	M	Frontal convexity	iso	iso	heterogeneous	(+)	(+)	(-)	(-)	
Present case		34	F	CP cistern				(+)	(-)	(-)	(-)	(-)

CP: cerebellopontine; NA: Not available; Calc: calcification; T1W1: T1-weigted image; T2W:1 T2-weigted image

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