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Review

Rare Lesions of the Cerebellopontine Angle

Serebellopontin Bölgenin Nadir Görülen Lezyonları

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ABSTRACT

Vestibular schwannomas, meningiomas and epidermoids account for a vast majority of the lesions occurring in the cerebellopontine angle (CPA). Neoplastic and non-neoplastic pathologies other than these tumors constitute 1% of all lesions located in the CPA. The aim of this study was to reveal our experience in the treatment of the rare lesions of the CPA. We have retrospectively reviewed the medical files and radiological data of all patients who underwent surgery involving any kind of pathology in the CPA. We have excluded those patients with a histopathological diagnosis of meningioma, schwannoma and epidermoids. Our research revealed a case of craniopharyngioma, a case of chloroma, a case of solitary fibrous tumor, a case of pinealoblastoma, a case of atypical teratoid rhabdoid tumor, a case of an aneurysm, a case of hemorrhage and a case of abscess.

KEYWORDS: Cerebellopontine angle, Posterior fossa, Tumor, Rare

ÖZ

Serebellopontin köşe tümörlerinin çok büyük bir kısmını vestibüler schwannomlar, meningiomlar ve epidermoid tümörler oluşturmaktadır. Bu patolojiler dışındaki neoplastik ve neoplastik olmayan lezyonların görülme yüzdesi yaklaşık %1 civarındadır. Biz de çalışmamızda serebellopontin bölge yerleşimli nadir lezyonlar üzerindeki tecrübemizi sunmak istedik. Serebellopontin köşe tümörü nedeniyle ameliyat ettiğimiz hastaların dosya ve filmleri taranmış, patoloji sonucu schwannom, meningiom ve epidermoid çıkan hastalar çalışma dışı bırakılmıştır. Serebellopontin köşe yerleşimli kraniyofarenjioma, kloroma, pinealoblastom, soliter fibröz tümör, atipik teratoid rabdoid tümör, anevrizma, hematom ve de abse olguları çalışmaya dahil edilmiştir.

ANAHTAR SÖZCÜKLER: Serebellopontin açı, Arka çukur, Tümör, Nadir

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INTRODUCTION

Cerebellopontine angle (CPA) tumors account for 5-10% of all intracranial tumors in adults and 1% of all pediatric intracranial tumors (14, 22, 26, 38). Among these, 80% are vestibular schwannomas followed by meningiomas and epidermoids (20, 23, 26, 32). A variety of unique lesions constitute less than 1% of all CPA tumors. Differential diagnosis of the pathologies located in the CPA is difficult due to the similarity of presenting signs and symptoms seen in various lesions. Advanced neuroradiological imaging can help to differentiate these rare lesions and to plan an appropriate treatment protocol.

The site of origin is the main factor in making a preoperative diagnosis for these rare tumors and attenuation at computed tomography (CT), signal intensity at magnetic resonance imaging (MRI), enhancement, shape, margins, extent, mass effect, and adjacent bone reaction help in the differential diagnosis (10).

This study presents a series of rare lesions of CPA that had been treated in our clinic. The history, neurological examination and radiological evaluation of each patient were reviewed. The surgery and prognosis of these patients are discussed.

MATERIAL and METHODS

Case 1

This patient was an 8-year old girl with a previous diagnosis of acute myeloblastic leukemia for which she had received chemotherapy. She was considered to be in remission when she was referred to us unconscious with a Glasgow coma scale (GCS) of 6. Cranial CT revealed a hyperdense lesion in the left CPA with a central necrotic area and MRI revealed a left extra-axial CPA lesion with dimensions of 4.5x4x3 cm. The lesion was isointense with gray matter on T1W (T1 weighted) images and minimally hyperintense on T2W (T2 weighted) images and was enhancing after intravenous gadolinium injection (Figure 1A,B,C). The fourth ventricle was compressed resulting in an obstructive hydrocephalus. MR spectroscopy (MRS) revealed an intralesional obvious choline peak with TE:135 and 270 msn values and choline/creatinine ratio was increased.

The patient was operated because of the life-threatening clinical condition and only subtotal decompression of the lesion could be achieved because of the high vascularity and massive bleeding of the tumor. Histopathological examination of the lesion revealed a necrotic tumoral infiltration within the cerebellum (Figure 2A). The tumor was composed of sheets of immature myeloblasts and included numerous areas of mitosis (Figure 2B). The cells showed eosinophilic cytoplasm and nuclear irregularity in focal areas. There were multiple tingible body macrophages among the neoplastic cells. Immunohistochemical analysis of myeloperoxidase displayed diffusely striking reactivity in the cytoplasm of the immature myeloblastic cells. The histopathologic diagnosis was consistent with granulocytic sarcoma. Unfortunately the patient did not show any clinical improvement and died on the sixteenth postoperative day (3).

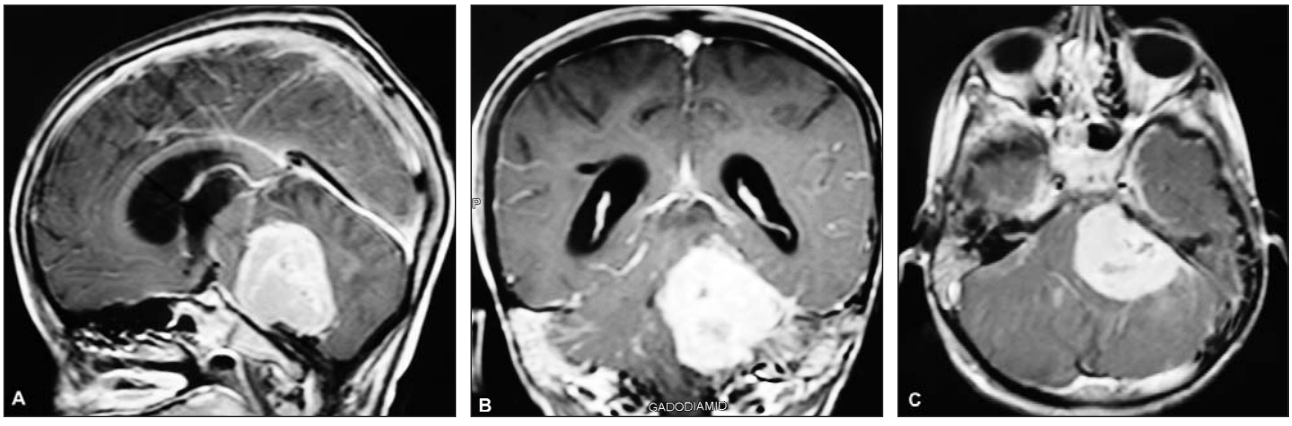

Figure 1: *T1-weighted postcontrast sagittal* ***(A)****, coronal* ***(B)*** *and axial* ***(C)*** *images showing an extra-axial mass lesion of the cerebellopontine angle with marked enhancement*

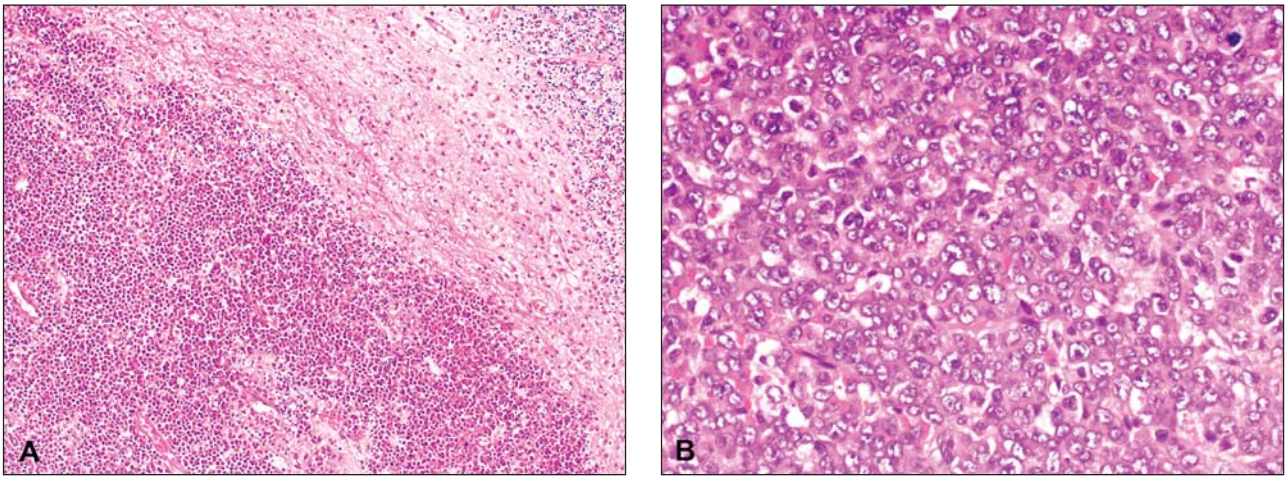

Figure 2: (A) *Sections included necrotic tumoral infiltration within the cerebellum (H&E x 100),* ***(B)*** *The tumor consisted of sheets of immature myeloid cells and included numerous areas of mitosis (H&E x 400).*

Case 2

A 56-year old male was admitted to our clinic with a history of imbalance for one month. Neurological examination was normal except slight impairment of cerebellar tests on the left. MRI revealed a 3x5x5 cm mass lesion in the left CPA. The mass was composed of both solid and cystic components and was enhancing heterogeneously following intravenous (iv) injection of gadolinium (Figure 3A,B). Preoperative radiological diagnosis was ependymoma or metastasis. The tumor was completely resected with a paramedian suboccipital approach. The tumor was seen to originate from the petrous dura and all cranial nerves were preserved. The postoperative course was uneventful and the patient was discharged from the hospital at the fourth postoperative day with a completely normal neurological examination.

Histopathological evaluation revealed a solitary fibrous tumor showing a fascicular proliferation pattern with intercellular collagen bundles (Figure 4A). They were composed of sheets of spindle and polygonal cells with oval nuclei and numerous slit-like vascular spaces occasionally showing a staghorn appearance. Mitotic figures were rarely seen. Special stains for reticulin highlighted a rich network invested around individual cells and in some areas around groups of cells (Figure 4B). Whorls, psammoma bodies and storiform cell arrangements, or necrosis were absent. Brain invasion was not observed. The Ki67/MIB-1 labeling index was 1.5%. Immunohistochemical analysis of the tumor cells demonstrated strong CD34 and vimentin reactivity and focal positivity for Bcl-2 and CD99 was observed (Figure 4C). The tumor did not express epithelial membrane antigen, and S–100 protein (36).

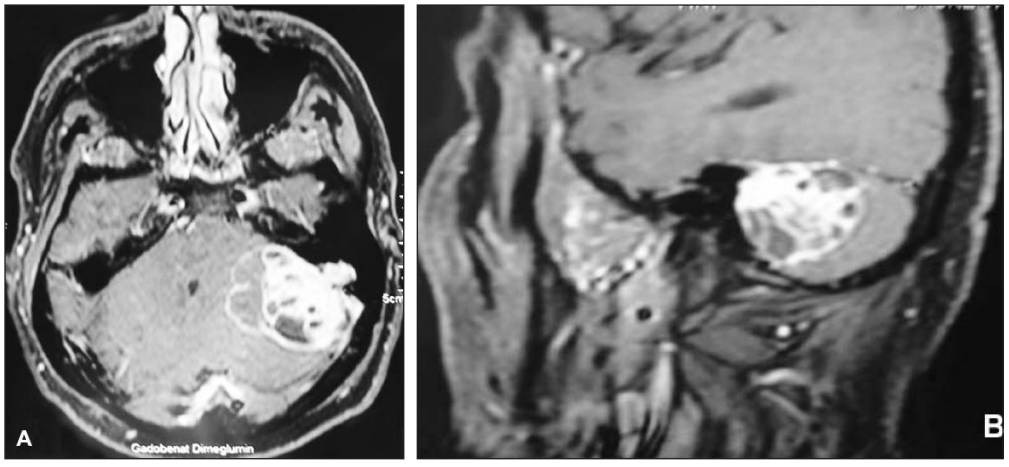

Figure 3: (A) *Axial T1-weighted cranial and* ***(B)*** *Sagittal T1-weighted cranial MRI showing 3 x 5x 5 cm mass lesion in the left cerebellopontine angle. The mass was composed of both solid and cystic components and enhanced heterogeneously after intravenous injection of gadolinium.*

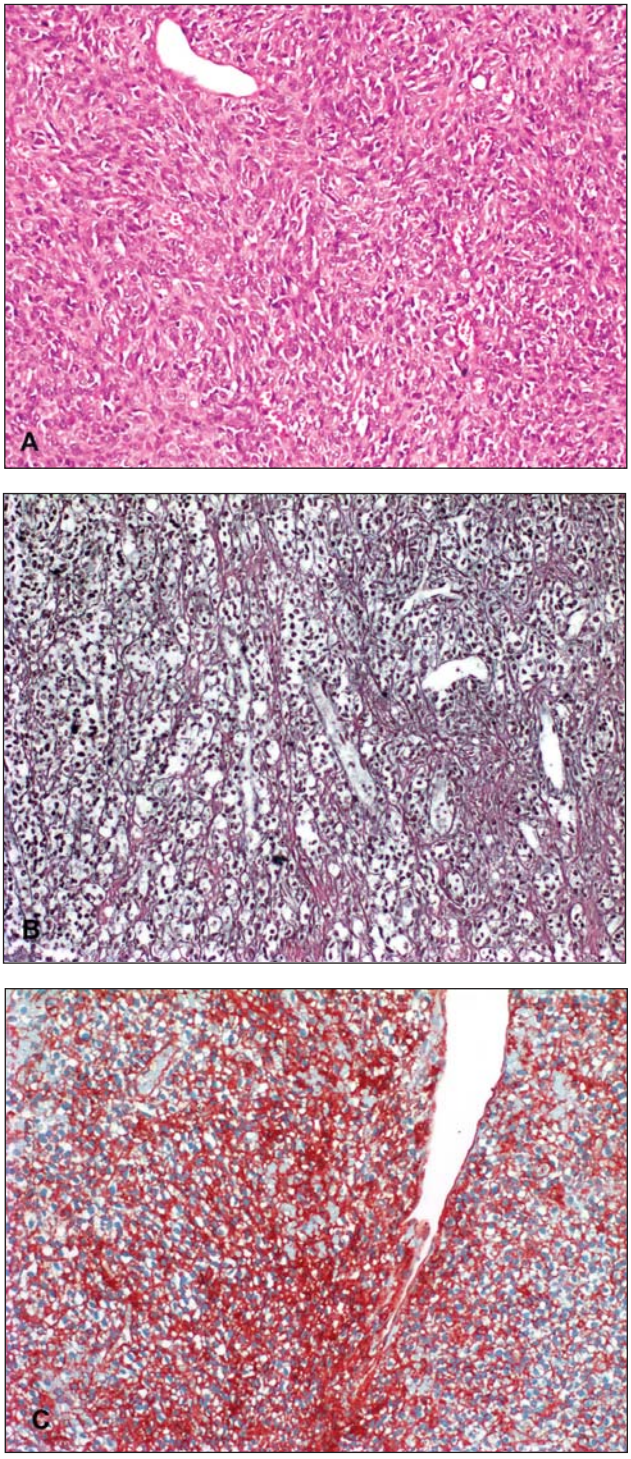

Figure 4: ***(A)*** *Meningeal solitary fibrous tumor with a fascicular proliferation pattern, which is associated with numerous cleft like vascular spaces (H&E x 100),* ***(B)*** *Silver staining showed a dense intercellular reticulin network (histochemistry x 200),* ***(C)*** *The tumor was strongly and uniformly immunoreactive for CD34 (immunoperoxidase x 200).*

The patient was followed up on a regular basis and second-year control examination was completely normal.

Case 3

A 14-year-old boy was admitted because of progressive hearing loss in the right ear for two years. He was also complaining of diplopia and neck pain radiating to shoulders. Neurological examination confirmed bilateral papiledema and sensory loss of trigeminal nerve. A slight peripheral paresis of the right facial nerve was a;sp noted and hearing was diminished on the right side. Computerized tomography (CT) showed a right cerebellopontine mass of 5x3x3 cm size (Figure 5). He was operated with a right paramedian incision and suboccipital craniectomy. The mass had a cystic component and macrocalcifications. The round and encapsulated tumor was removed totally with preservation of lower cranial nerves. Histopathological examination of the mass revealed a diagnosis of craniopharyngioma (2).

Case 4

A 2-year-old girl was admitted to our hospital with a 1-week history of a cervical tilt to the left side. Her physical examination was normal except for the cervical tilt and neurological examination was normal. Cranial MRI and cranial CT revealed a cystic mass of 3x3x2 cm size in the right cerebellopontine angle that was enhancing heterogeneously following iv godolinium infusion. The brainstem was pushed

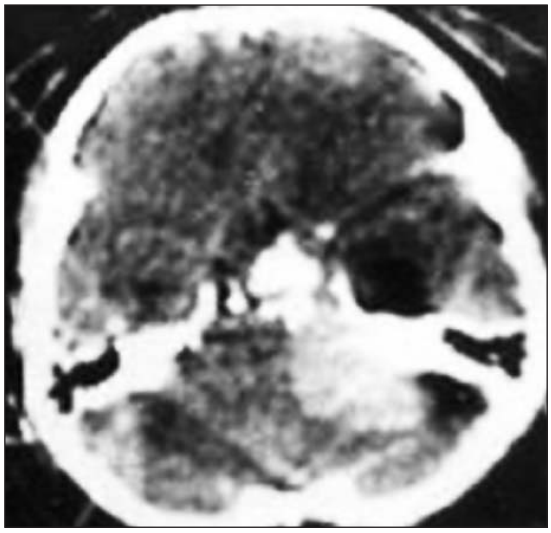

Figure 5: *Axial cranial CT showing a large mass lesion located on the left CPA.*

to left by the mass and the border of the mass and brainstem seemed blurred giving the impression of an infiltrative characterization of the mass (Figure 6A,B).

The patient was operated and the mass was partially resected preserving the cranial nerves. Postoperative period was complicated with left-sided sixth nerve palsy and right-sided central facial paralysis. She was referred to the department of oncology for chemotherapy and radiotherapy.

The histopathological examination of the tumor yielded a diagnosis of atypical teratoid rhabdoid tumor which is the combination of small undifferentiated cells and large pale cells with rhabdoid characteristics and a characteristic immunohistochemical profile that is the tumor revealed positivity for neuron-specific enolase, smooth muscle actin, and epithelial membrane antigen (15).

Besides these cases, we also experienced a case of pinealoblastoma, a case of an aneurysm, a case of hemorrhage and a case of abscess in the cerebellopontine angle (Figure 7A,B,C,D).

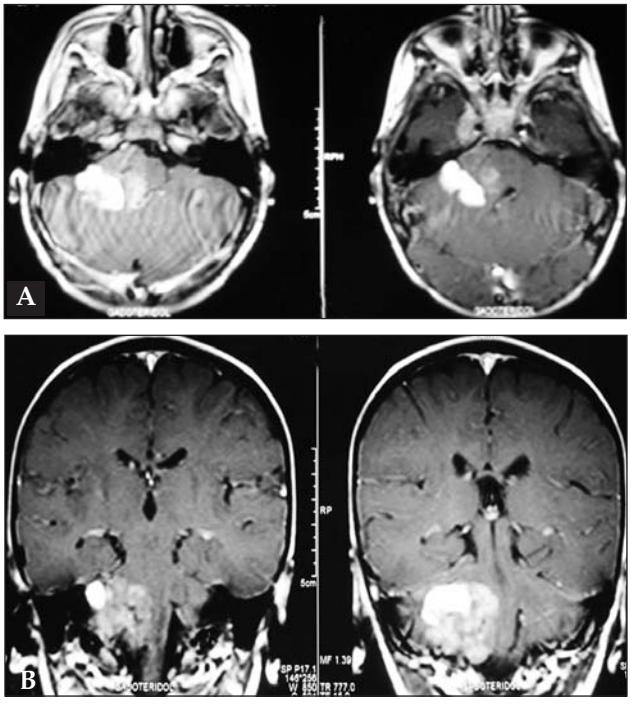

Figure 6: ***(A)*** *Axial T1 MRI scan after a contrast injection showing an extensive irregular mass at the right cerebellopontine angle and the infiltration of the tumor into the brainstem* ***(B)*** *Coronal T1 MRI scan after a contrast injection showing an extensive irregular mass at the right cerebellopontine angle and the infiltration of the tumor into the brainstem.*

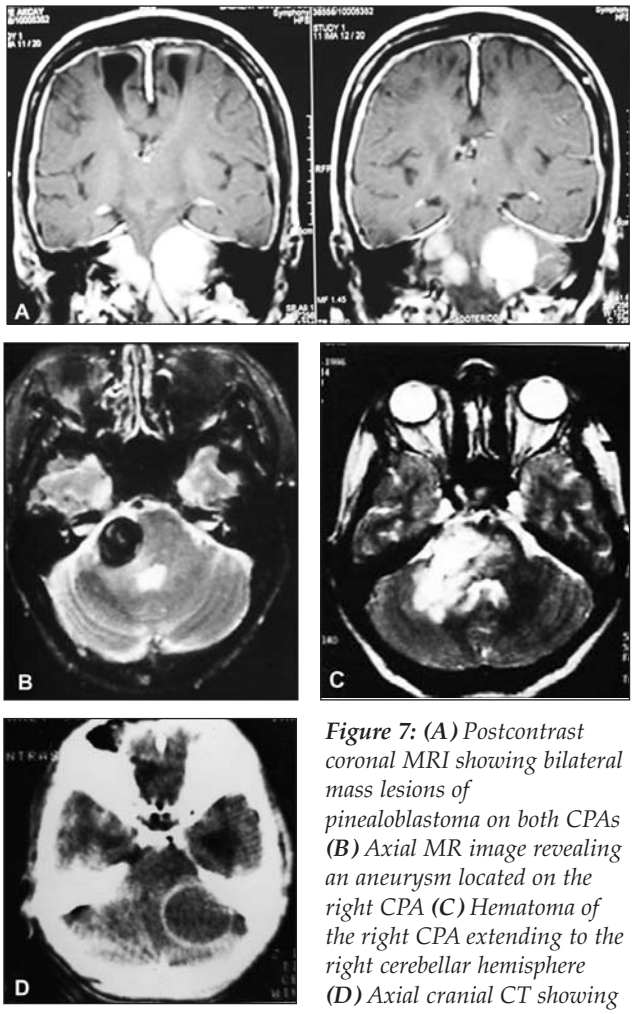

Figure 7: ***(A)*** *Postcontrast coronal MRI showing bilateral mass lesions of pinealoblastoma on both CPAs* ***(B)*** *Axial MR image revealing an aneurysm located on the right CPA* ***(C)*** *Hematoma of the right CPA extending to the right cerebellar hemisphere* ***(D)*** *Axial cranial CT showing an abscess on the left CPA.*

DISCUSSION

The term cerebellopontine angle was first introduced in 1902 by Henneberg and Koch when they reported two individuals with bilateral acoustic neuromas (ANs) occurring in the location they described (in German) as the kleinhirnbruchen-winkel (kleinhirn= cerebellum; bruchen = pons or bridge; and winkel = angle) (16). Surgery of the pathologies located in CPA had long been associated with a considerable risk of mortality and morbidity due to the complex structure of this area and its close proximity to cranial nerves and vessels. Refinements in neuroanesthesia, achievements of surgical approaches and microneurosurgical techniques have improved surgical results to acceptable mortality and

morbidity risk level. Superior neuroradiological techniques contribute to this improvement by both helping the differential diagnosis of these pathologies and also guiding the surgeon in the selection of the most appropriate operative approach.

MRI with gadolinium diethylenetriaminepentaacetic enhancement is the gold standard diagnostic tool and enables detection of tumors as small as 2-3 mm.(10, 35). The site of origin and differential signal characteristics of MRI can help to differentiate between unusual tumor types.

Lesions in the CPA may be categorized into three main groups: 1) lesions originating in the CPA, 2) lesions primarily located in close anatomical sites and extending to the CPA and 3) intraventricular and brain stem pathologies showing exophytic expansion to the CPA (6). Vestibular schwannomas, and meningiomas constitute the first group and compose a very high percentage of the CPA lesions (20, 23, 26, 32). Arachnoid cysts, nonacoustic schwannomas, aneurysms, melanomas and embryologic remnants like epidermoid cysts, dermoid cysts, lipomas also arise primarily from the CPA (28, 37). Cholesterol granulomas, paragangliomas, chondromatous tumors, chordomas, endolymphatic sac tumors, pituitary adenomas, and apex petrositis are the pathologies that had been reported to extend to CPA from close anatomical sites (11, 14). Astrocytomas, choroid plexus papillomas, lymphomas, hemangioblastomas, ependymomas, medulloblastomas, and dysembryoplastic neuroepithelial tumors are the tumors that have been reported to invade CPA secondary to exophytic extension (18).

Vestibular schwannomas are isointense to hypointense to brain on T1-weighted images and hyperintense on T2-weighted images and enhance after iv contrast. They may be heterogenous due to cystic degeneration, hemorrhage or vascularity. They are generally round or oval in their cisternal portion and taper along the axis of the internal acoustic meatus (37). Meningiomas are usually isointense to brain on T1-weighted images, can be isointense, hypointense or hyperintense on T2-weighted images. They enhance strongly and homogeneously with contrast and a dural tail of enhancement may aso be present. They do not show extension into the internal acoustic meatus and usually have a broad base against petrous bone. Epidermoids may be more commonly hypointense on T1-weighted images, hyperintense on T2-weighted images and slightly brighter than cerebrospinal fluid on intermediate images reflecting the keratomatous debris and cholesterol in them. Less commonly they may be hyperintense on T1-weighted images, hypointense on T2-weighted images reflecting their high lipid content (37).

In our cases of aneurysm, hemorrhage and abscess, the diagnosis was easy on a radiological basis. The case of chloroma had a medical history that helped in the diagnosis. In the cases of craniopharyngioma, SFT, atypical teratoid rhabdoid tumor and pinealoblastoma, the radiological features were not suggestive of the pathology.

Chloromas are extramedullary leukemias which can be encountered at any anatomical location, the common sites being the orbit, skin, bones, paranasal sinuses and epidural areas (13,33). Only three cases have previously been reported in the CPA and our case is the fourth case. To the best of our knowledge, no more cases have been reported.

Solitary Fibrous Tumors (SFT) are rare spindle-cell neoplasms of mesenchmal origin first described by Klemperer and Robin in the visceral pleura in 1931 (21). Since then, they have been described in many extra-pleural sites including the central nervous system (CNS). Primary meningeal SFT was first described by Carneiro et al. in 1996 and to our knowledge less than 100 SFTs have been reported both in the cranial and the spinal compartments of the CNS between 1996 and 2008 (4, 8, 25, 27, 29, 34). Among these, four were located in the cerebellopontine angle and our patient is the fifth case.

Craniopharyngiomas account for 1.2 to 4% of all intracranial tumors and typically develop in the infundibulohypophysial axis (1, 7). They are thought to derive from remnants of the Rathke pouch epithelium and typically occupy the sella and/or the suprasellar cistern. Rarely they had been reported to be located in the optic nerves, the pineal region, the sphenoid bone, the pharynx, and the cerebellopontine angle which can be either the primary location or extension of a suprasellar tumor (2, 9, 12, 24). Our case is the first reported case of cerebellopontine angle craniopharyngioma and a few more cases have been reported up to date (9, 12, 24).

Atypical teratoid/rhabdoid tumors have been wrongly diagnosed in the past as medulloblastomas and are highly malignant lesions of childhood that carry a very poor prognosis. Biggs and colleagues were the first to describe an intracranial malignant rhabdoid tumor, and Rorke and colleagues referred to a primary malignant rhabdoid tumor of the central nervous system as an "atypical teratoid / rhabdoid tumor" (ATT/RhT) because it consisted of a unique combination of neuroepithelial, peripheral epithelial, and mesenchymal elements (5, 17, 30). Rorke and colleagues also found that ATT/RhTs developed at the cerebellopontine angle in 15% of the subjects in their series (30, 31).

CONCLUSION

Our experience has suggested that lesions considered rare may occur more frequently than expected in CPA. Maintaining a high index of suspicion and careful analysis of preoperative MRI images can help in the differential diagnosis and guide in planning the most appropriate treatment protocol for these pathologies.

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