Subependymoma of the Fourth Ventricle with an Unusual Presentation

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ABSTRACT

We present a rare case of a 62 year old female patient with a left tonsillar cerebellar tumor that presented with migraine headaches, left facial numbness and dysphasia. Brain Magnetic Resonance imaging (MRI) showed a small enhancing lesion in the inferior aspect of the fourth ventricle without hydrocephalus. After craniotomy and subtotal excision of the tumor, histological examination of the specimen showed clusters of tumor cells with isomorphic nuclei embedded in a dense fibrillary matrix with microcystic changes and calcifications, corresponding to a subependymoma. These tumors are benign and correspond to WHO grade I with an overall good prognosis. They represent less than 1% of all intracranial tumors and are more common in males. We describe the clinical and radiological findings as well as the histologic appearance in this case.

INTRODUCTION

Subependymomas are benign tumors composed of clusters of ependymal and astrocyte-like cells corresponding to WHO grade I. They represent less than 1% of all intracranial tumors, and 8% of ependymal tumors. Subependymomas are usually more common in middle-aged patients and elderly, with a higher incidence in men; but, they can occur at any age. They usually present more commonly in the fourth ventricle, followed by lateral ventricles; although, they can also be found in the cerebral or cervicothoracic medulla, third ventricle and septum pellucidum. Subependymomas are usually asymptomatic, but they can become clinically apparent if ventricular obstruction occurs or if they present in the spinal cord. In this report, we present a patient with an uncommon clinical presentation of a subependymoma in the fourth ventricle, describing its clinical, radiological and histological findings.

CASE REPORT

This is a 62 years old female patient with history of diabetes mellitus type 2 and hypertension that presented with migraine-type headaches. In addition, she complained of abysmal left facial pain and dysphasia that were described as recurring numbness and burning sensations. The patient denied nausea, vomiting and weight loss and presented with an unremarkable physical and neurological evaluation. A Brain MRI performed (fig.1) showed an approximately 8mm x 6mm x 6mm midline enhancing mass located in the inferior aspect of the fourth ventricle without producing hydrocephalus but exerting a mild mass effect on the dorsal medulla. The mass was mildly hyperintense on FLAIR and T2-weighted images, but isointense in T1-weighted images with increased enhancement after gadolinium-based contrast administration. The patient underwent a suboccipital craniotomy and a left tonsillomedullary and fourth ventricular subependymoma (fig. 2). Immunohistochemical stains illustrated a strong positivity for S-100 (fig1B) and glial fibrillary acidic protein (GFAP) (fig 3D) and with a very low proliferative index (1-2%), as demonstrated by Ki67 (fig1C).

Fig 1. MRI features of subependymoma [A]: MRI-FLAIR; [B]: T1-weighted with contrast

Fig 2. Histopathology of the mass. [A]: Hematoxylin and eosin stain (H&E) of clusters of nuclei in a dense fibrillary stroma (10x); [B]: Cystic degeneration in H&E (20x); [C]: Calcifications in H&E (40x)

DISCUSSION

Subependymomas usually remain asymptomatic; but, can progress to a clinically apparent disease. The most common presenting symptoms frequently occur due to an obstruction of the ventricular system and are correlated with tumor size. A study of 83 cases showed that the most common symptoms were associated with an increased intracranial pressure, such as headaches, gait problems, ataxia, nausea, dizziness and vomiting[1]. Other clinical reports mentioned seizures, sensory symptoms and motor weakness, primarily caused by compression of neural structures[2]. The latter, is rare presentation of this disease. To our knowledge, there has been one case reported with symptoms of sensory dysesthesias that included the face[2]. The patient in our report had an unusual presentation of sensory dysesthesias on the left side of the face and headaches not associated with hydrocephalus.

MRI characteristics of the lesion can be very useful in defining the site and extent of the tumor, as well as its association to surrounding structures and can help in pre-operative planning[3]. On the other hand, the features that are most commonly associated to subependymomas have not been shown to be pathognomonic due to its variability. On the MRI of this case report, the lesion was described as mildly increased intensity relative to brain parenchyma and increased enhancement with contrast in T1-weighted images. Many reports show that most of these lesions are hypo- to isointense on T2-weighted imaging and hyperintense on T2-weighted images, which differs with our T2-weighted image on the lesion. Furthermore, reports show variability in contrast-enhanced images; they are most often absent, but may show mild to intense enhancement[1,2,3,4]. Therefore, the radiographic features in this case demonstrate the variability that can be observed among this type of tumor.

The pathological findings of the lesion correspond to a subependymoma of the fourth ventricle. Histological analyses showed clusters of small nuclei separated by a fibrillary matrix of glial cell processes. Also present were: calcifications, cystic degeneration, vascular hyalinization, pleomorphic and mild atypia. Absence of mitosis, necrosis, perivascular pseudosarcomatous or true rosettes were noted. The lesion was strongly immunoreactive to S-100 protein and GFAP which indicated its astrocytic differentiation. In addition, the tumor had a very low Ki-67 proliferation index (1-2%). Literature review suggests a correlation of increased Ki-67 with tumor recurrence[4,5], and clinical behavior[6]. A study of 83 cases of subependymomas showed that an abrupt transition and vascular hyalinization were the most common histopathological features; nonetheless, pleomorphism, cystic degeneration, mixed histology, calcification, hemorrhage, necrosis and mitosis may also be observed in descending incidence[7]. Another report described a rare subependymoma with prominent Rosenthal fibers[8]

CONCLUSIONS

- Subependymomas are an unusual type of ependymal tumor presenting most frequently in middle aged and elderly patients and more than twice as frequent in men than women.
- There is a great variability of clinical presentations and, most of the time, symptoms are related to obstructive hydrocephalus.
- Radiologic correlation with identification of a lesion growing within the ventricular system is of great diagnostic value.

BIBLIOGRAPHY

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