Case Report

Intracerebral Schwannoma: A Case Report

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Summary

Schwannomas are encapsulated, benign tumors, which generally originate from the schwann cells on cranial or peripheral nerves. Malignant transformation into neurofibrosarcoma occurs in 1% of these tumors. While schwannomas account for 8% of all central nervous system tumors, intracerebral schwannomas occurring independent from cranial nerves are extremely rare. Their treatment is usually surgical and total excision is generally curative. In this case report, we present a 27-year-old male patient who was admitted to our clinic with focal motor seizures due to a left frontal mass demonstrated on radiological imaging. Surgical treatment was planned and total excision was achieved. Histopathological investigations of the resected mass showed an extremely rare entity of intracerebral schwannoma. Intracerebral schwannomas are further discussed in light of the relevant literature.

Key words: Intracerebral schwannoma, schwannoma, S100, GFAP, calretinin, CD56

INTRODUCTION

Schwannomas are also known as neurolemmoma, neurinoma and Schwann cell tumors. They originate from Schwann sheath cells. Schwannomas are homogenous tumors that contain only Schwann cells. They comprise 8% of all central nervous system tumors and they most commonly involve vestibular portion of the 8th cranial nerve. Their incidence shows a 2:1 male predilection and the rate...
of malignant transformation is less than 1% in all schwannoma cases. They may originate from any cranial nerve except for the optic nerve. They often manifest as a part of neurofibromatosis type 1 (von Recklinghausen disease). On immunohistochemical studies, eosin-hematoxylin staining may be enough for a diagnosis. However, a differential diagnosis may be required and they can always be differentiated from other tumors by their positive reactions to S-100 and reticulin stains.

Intraparenchymal involvement of schwannomas is extremely rare. First appearance of intraparenchymal schwannomas in the literature was reported by David et al in 1965 and less than 50 cases have been published up until today. Reported cases in the literature demonstrate that intracerebral schwannomas tend to occur in the first two decades of life. The recurrence rate is very low and total surgical removal is the definitive treatment.

In this case report we present a rare case of an intraparenchymal schwannoma and provide the histopathological findings.

CASE PRESENTATION

A 27 years old male patient presented at our clinic with focal motor epileptic attacks that had been becoming more frequent over the past year. His neurological examination revealed no neurological deficits. No speech abnormalities were noted. A cranial MRI with IV gadolinium injection was obtained. A homogeneously enhancing mass lesion with the dimensions of 3x2x2 cm was found in the left frontal lobe. The lesion mostly involved the middle frontal gyrus with minimal edema involving the frontal operculum on T2W images. Glioma and meningioma were considered as the most likely diagnoses. Surgical treatment for tumor removal was planned. Left frontal craniotomy was performed. Gross total resection of the tumor was achieved via middle frontal gyrus corticotomy. Intraoperatively, the tumor had a yellow-to red coloring with a hard circumscribed texture. Minimal edema was noted reaching the frontal operculum. The recovery was uneventful. Postoperatively, no speech disorder was noted or reported by the patient ever since. Valproic acid was continued and the patient remained seizure-free.

The pathological examination revealed a well-demarcated tumor from adjacent neural tissue containing a constellation of nodules of different sizes and shapes. The tumor consisted of monomorphic serpentine cells forming intertwining fascicles. The cells typically displayed oval to spindle nuclei with granular chromatin and a fibrillated cytoplasm, which were consistent with Schwann cells in an Antoni A region in a Schwannoma. The Ki 67 index was reported as 1%. Conclusively, the tumor was diagnosed as Schwannoma grade I.
**Figure 1:** Preoperative (A, B) and postoperative MRI (C, D)

**Figure 2:**
- Fig 2a. Well demarcated tumor from adjacent neural tissue. (HE, x200)
- Fig 2b. Intertwining fascicles formed by spindle cells. (HE, x200)
- Fig 2c. Parallel bundles of Schwann cells. (HE, x400)
- Fig 2d. Characteristic reticulin framework of Schwannoma. (Gomori reticulum stain, x200)
- Fig 2e. S100 reactive cells forming fascicles. (Biotin streptavidin complex (ABC), x400)
- Fig 2f. Schwann cells showing intense staining with GFAP. (Biotin streptavidin complex (ABC), x400)
- Fig 2g. Calretinin reactive cells of the neoplasm. (Biotin streptavidin complex (ABC), x400)
- Fig 2h. CD56 reactive cells of the neoplasm. (Biotin streptavidin complex (ABC), x400)
- Fig 2i. Mast cells showing specific tryptase immunoreactivity with MCT. (Biotin streptavidin complex (ABC), x400)
DISCUSSION

Intracranial Schwannomas constitute 8% of all primary brain tumors and the overwhelming majority involves the 8th cranial nerve\textsuperscript{(16)}. Seventy-eight % of Schwannomas occur in the cerebellopontine angle\textsuperscript{(15)}, and the intraparenchymal Schwannomas are extremely rare. The literature includes reports of Schwannomas located in medulla oblongata\textsuperscript{(1,14)}, in the ventricles\textsuperscript{(5)}, sellar region\textsuperscript{(10)} and in the supratentorial region with both intra and extra-axial locations\textsuperscript{(6,12)}. In our case, the tumor was located in the left frontal area.

Clinical presentation will be determined by the location of the tumor. Headache, seizures and focal neurological deficits are the most common symptoms\textsuperscript{(1,6,8,17,18,22)}. For the schwannomas with typical locations, the radiological studies may reveal characteristic signs such as isodense or hypodense masses on CT imaging. Similarly, MR imaging may reveal lesions with hypointensity on T1W and iso- or hyperintensity on T2W imaging with heterogeneous contrast enhancement after gadolinium injection. Our patient had an isointense lesion on T1W images and heterogeneous contrast enhancement after contrast injection (Fig 1).

A few hypotheses have been proposed to explain the intraparenchymal development of these tumors with apparent nerve sheath cell origin. These Schwannomas have been suggested to grow from the Schwann cells in the perivascular nervous plexus\textsuperscript{(9,13)} or by transformation of the primitive multi-potent mesenchymal cells into Schwann cells\textsuperscript{(7)}. They were also proposed to result from a migration defect of the neural crest cells during central nervous system development\textsuperscript{(16,20)}.
Radiological and pathological differential diagnosis for these tumors includes pilocytic astrocytoma, lymphoma, pleomorphic xanthoastrocytoma, meningioma, ganglioglioma, metastasis and solitary fibrous tumor (SFT)\(^{(3,12,23)}\). During the histological differential diagnosis, fibrillary astrocytoma was ruled out with IDH negativity, solitary fibrous tumor was ruled out with CD34 negativity, fibrous meningioma was ruled out by showing lack of expression for progesterone receptor and epithelial membrane antigen (EMA). A previously proposed immunoreactivity chart for the differential diagnosis of schwannomas, neurofibromas and some other common cerebral tumors is presented in Table 1\(^{(25)}\).

The diagnosis of intracerebral schwannoma was based on S100, MCT, GFAP, CD56 as well as calretinin positivity (Fig 2, Table 1). Calretinin and CD56 positivity have been previously proposed to differentiate schwannomas from neurofibromas, which may also stain with S100\(^{(24,25)}\). As in our case, immunohistochemical studies are generally sufficient for a definitive diagnosis but further investigation may be conducted via electron microscopy\(^{(21)}\).

In conclusion, intracerebral schwannomas are rare, benign tumors. Pathological investigation is required for a definitive diagnosis. Surgical total resection is curative.

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