Case Report

Solitary fibrous tumor of the central nervous system: report of 2 cases and review of literature

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Abstract: Solitary fibrous tumors (SFTs) rarely occur in the central nervous system (CNS). Involvement of the brainstem and pineal gland is rarely recorded. Herein, we represent 2 cases of SFTs and firstly report SFT of the pineal gland. Cranial MR imaging showed isointense to hypointense signal intensity, and marked enhancement. Microscopically, the tumors showed characteristic “patternless-pattern” architecture. Elongated tumour cells formed fascicles alternating with hypocellular densely collagenous stroma. Immunohistochemistry for CD34, BCL2, and CD99 favors the definitive diagnosis of SFT. It is difficult to predict prognosis in patients with intraventricular SFT. In general, complete surgical resection may offer the best chance of a favorable clinical outcome.

Keywords: Solitary fibrous tumor, central nervous system, immunohistochemistry, magnetic resonance imaging, CD34

Introduction

Solitary fibrous tumors (SFTs), rare mesenchymal spindle cell neoplasms, are frequently arising from the pleural cavity. It rarely occurs in extrapleural sites like upper respiratory tract, lung, nasal cavity, paranasal sinuses, orbits, mediastinum, major salivary glands, breast, meninges, liver and urogenital organs [1, 2]. The central nervous system (CNS) SFT was firstly described by Carneiro et al. in 1996 [3]. Approximately a hundred and odd cases have been reported previously at various sites within the CNS. Involvement of the brainstem and pineal gland is rarely recorded. Herein, we represent 2 cases of SFTs and firstly report SFT of the pineal gland: we discuss their clinical, imaging feature, histological features, and differential diagnosis.

Case report

Case one

A 44-year-old female presented with a five-day history of headache and dizziness. Her neurologic examination was normal but MRI demonstrated a 2.2×2.5×2.1 cm round-like mass with clear margin in pineal region. Solid portions of mass showed low signal on T1WI (Figure 1A) and high signal on T2WI (Figure 1B) with markedly enhancement on contrast, while cystic portions presented as multiple cystiform long T1 and long T2 signal intensity, not enhanced on contrast. Adjacent brain tissue was lamellar long T1 and slightly long T2 signal intensity meaning edema and markedly enhanced on contrast (Figure 1C). Imaging findings suggested a germinoma. The patient underwent pineal region lumpectomy.

Pathologic examination of the tumor revealed a spindle cell tumor with a “patternless-pattern” and areas of dense collagen deposition. Tumor composed of cellular and paucicellular areas. In the cellular areas, the tumor showed interlacing fascicles of spindleshaped cells with moderate amount of cytoplasm and oval to elongated nuclei exhibiting variable pleomorphism (Figure 2A). No mitotic activity was noted. In the paucicellular areas, dense bands of collagen were seen separating the cells (Figure 2B). The stroma was
myxoid in areas and thin vascular channels were present. Immunohistochemically, tumor cells showed strongly positive for vimentin and CD34, weakly positive for CD99, partially positive for Bcl-2 and S-100 and negative for EMA (Figure 2C). Ki-67 immunostaining showed a 5% proliferative index. The patients made an excellent recovery with a normal examination and no evidence of recurrence six months after surgery.

Case two

A 52-year-old male presented with numbness and weakness in all four extremities for one week. Physical examination showed hypalgesia, hypothermesthesia and hypopselaphesia of the whole body. Brain MRI revealed a 2.3×1.8×2.7 cm fusiform mass with clear margin in front of the medulla oblongata, was showed isointense and low signal on both T1WI (Figure 1D) and T2WI (Figure 1E). It was markedly heterogeneous enhanced after enhancement, in which emerged dot-strip non-enhancement, and the meninges in front of cervical spinal cord were markedly enhanced so that the lesion was separated from the medulla oblongata (Figure 1F). The smaller medulla oblongata was compressed and displaced to the right and backward. The imaging appearance raised a differential diagnosis including meningioma and neurilemmoma. The patient underwent brainstem lumpectomy.

In the histopathological examination, pattern variation was observed. Elongated tumour cells formed fascicles alternating with hypocellular densely collagenous stroma, similar to morpho-
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Immunohistochemically, the tumor cells were strongly positive for vimentin and CD34, weakly positive for S-100 and negative for EMA (Figure 2F). Ki-67 was expressed by less than 1% of cells. He has been lost to follow-up from three months after operation.

Discussion

SFT is a relatively new entity and our understanding of its nature is still limited. Although pleural SFT is still the most common, SFT has been described in various extrapleural sites, including soft tissues, the abdominal cavity, and the CNS [4]. SFTs can present in various locations with the CNS [5-8] and can be metastatic to visceral organs [9-11]. Herein, we reported 2 cases of SFTs in the CNS. To our best knowledge, SFT of the pineal gland has not been previously described.

The classic histologic features combined with immunohistochemistry are helpful in reaching a correct diagnosis. The characteristic histologic features of SFT include the so-called “patternless-pattern” of spindle cells, hemangiopericytoma-like pattern of vascularity, and thick strands of stromal collagen. Alternating hypo- and hypercellular areas is another important clue on low-power examination. Intense immunoreactivity for CD34 and Bcl-2 favors the diagnosis of SFT. Hypo- and hypercellular areas, collagenized stroma and strong CD34 positivity favored the diagnosis of SFT in the present case.

SFTs are difficult to distinguish from hemangiopericytoma and the general consensus in the nonurologic community is to combine those 2 entities into 1 entity. In the nervous system, however, the World Health Organization committee on the classification of tumors still maintains that those entities are separate. Hemangiopericytomas (HPCs) are remarkable for their characteristic thin-walled, branching or “staghorn” blood vessels. Although cellularity is variable, HPCs are often composed of closely packed, randomly oriented cells with irregular, carrot-shaped nuclei. The CD34 staining for SFT is usually strong and diffuse, whereas that of hemangiopericytoma is inconsistent and patchy. No specific molecular markers are known so far for SFTs. In contrast to soft tissue SFTs, within the CNS, “HPCs” are recognized as aggressive tumors with high rates of recurrence and metastasis [12]. However, Corinne et al. found overlapping pathological features and

Figure 2. Histopathological and immunohistochemical images of two cases. (A & B) The tumor of case two showed a spindle cell tumor with a "patternless-pattern" and areas of dense collagen deposition. Tumor composed of cellular (A) and paucicellular (B) areas. (C) Tumor cells showed strongly positive for CD34. (D & E) Elongated tumor cells formed fascicles alternating with hypocellular densely collagenous stroma. (F) Tumor cells were strongly positive for CD34.
common prognostic factors between SFT and HPC, suggesting that they belong to the same spectrum of tumors [13]. It is difficult to predict prognosis in patients with intraventricular SFT given the relative rarity of the lesion in the CNS, particularly at this site, and the limited information available regarding long-term outcomes. Metellus et al. showed a 50% recurrence rate over a median follow-up period of 45 months with a statistically significant association to incomplete surgical resection [14]. Histological features such as necrosis, hypercellularity and mitoses did not affect the rate of recurrence. A high proliferation index should be considered as a prognostic parameter. Michele et al. advocated inclusion of high Ki-67 (i.e., >5%) as an adverse prognostic parameter in assessing the prognosis of SFT of the CNS [15]. In the study, the tumor of case two showed hypercellularity and high Ki-67 (10%) index. There have still been no recorded recurrences. Therefore, in keeping with SFT at other sites in the CNS, complete surgical resection may offer the best chance of a favorable clinical outcome.

In conclusion, SFTs are rare in the central nervous system, so this diagnosis is usually overlooked by physicians. Careful radiological and histopathological examination is required to arrive at an accurate diagnosis. Total tumour removal can usually be achieved with an appropriate surgical technique.

Disclosure of conflict of interest

None.

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References

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