Abstract

Background: Rhabdomyosarcoma is a malignant tumor of the skeletal muscle cells. Those tumors mainly affect children and arise in primitive fetal mesenchyme even at sites that do not contain skeletal muscle. However, very few intracranial rhabdomyosarcomas have been reported. No previous case of primary intrasellar rhabdomyosarcoma except for 1 case has been reported in the literature.

Case Description: A 44-year-old woman presented with fatigue and diplopia associated with amenorrhea and galactorrhea. Visual field evaluation revealed a superior temporal deficit in both eyes. A right afferent pupillary defect was present. Magnetic resonance imaging revealed an intrasellar tumor with suprasellar extension. The tumor involved the sella turcica and right cavernous sinus. The patient was preoperatively diagnosed as pituitary adenoma, which was confirmed pathologically as rhabdomyosarcoma after the operation.

Conclusions: Our findings suggest that rhabdomyosarcoma should be considered in the differential diagnosis of a primary intrasellar neoplasm.

1. Introduction

Rhabdomyosarcoma is a malignant tumor of the skeletal muscle cells, which mainly affects children and occurs predominantly in the regions of the head and neck, followed by the genitourinary tract, retroperitoneum, and extremities. It occurs in primitive fetal mesenchyme even at sites that do not contain skeletal muscle. In the head and neck region, the orbit, cranium, nasal cavity, and nasopharynx are common sites of origin [17]. However, few intracranial rhabdomyosarcomas not accompanied by lesions at other sites has been reported [1]. We report a case of a rhabdomyosarcoma occurring within the sella turcica, which had been misdiagnosed as pituitary adenoma.

2. Case history

A 44-year-old woman presented with 1 month of fatigue and diplopia associated with 3 months of amenorrhea and galactorrhea. Her visual deterioration affected the right eye more than the left. On examination, she looked alert and lucid. Visual field evaluation revealed a superior temporal deficit in both eyes. A right afferent pupillary defect was present. She was unable to abduct her right eyeball. She could count fingers in the nasal field within the distance of 15 cm using her right eye. Her left eye had a visual acuity of 0.4. The remainder of her neurologic and general physical examination yielded normal results. Laboratory tests for pituitary hormones were submitted preoperatively; the results became available postoperatively and were within the reference range, with the exception of an elevated prolactin level of 1102 MIU/L (reference range, 66-721 MIU/L).

Magnetic resonance imaging (MRI) revealed an intrasellar tumor, about 2.5 cm in diameter, with suprasellar
The tumor involved the sella turcica and right cavernous sinus with intact inferior floor of the sella (Fig. 1). Compared to the surrounding brain tissue, the tumor was hypointense on T1-weighted images (Fig. 1A). It was heterogeneously enhanced with gadolinium (Fig. 1B and C). No abnormalities were found in the nasopharynx, nasal sinuses, paranasal sinuses, or brain parenchyma. Therefore, a diagnosis of pituitary adenoma had been assumed preoperatively.

To obtain an optimum decompression of the optical nerve, we chose a transfrontal approach to remove the tumor. In the operation, a reddish and soft tumor with a mildly thick membrane was found in the sellar region. It encroached the right cavernous sinus laterally and pushed the chiasm upward. The tumor was resected in piecemeal extension.

Fig. 1. Preoperative MR images depicting a mass in the sella turcica with suprasellar and right cavernous sinus extension. The tumor was visualized hypointense against gray matter on T1-weighted imaging (A) and heterogeneously enhanced with gadolinium (B, C).

Fig. 2. One month postoperative T1-weighted MR images after gadolinium enhancement showing a marked enlargement of the tumor (T). The tumor extended peripherally and protruded into the right frontal lobe. (A) axial image; (B) sagittal image.
fashion. The residual membrane was treated with coagulations. After gross total removal of the tumor, the pituitary gland was visualized in the left corner of the sella turcica. With those intraoperative manifestations, a clinical diagnosis of pituitary adenoma had been believed.

Three days after the surgery, the patient contracted diabetes insipidus, which lasted for more than a month followed by high fever. Because the patient’s situation continued to worsen, she subsequently underwent gadolinium-enhanced MRI scan again. The MRI showed a tumor even bigger than the previous one in the same site, which extended peripherally and protruded into the right frontal lobe of the parenchyma (Fig. 2). Accordingly, the patient underwent another procedure. This time, the tumor was found to be elastic and fibrous. It seemed more infiltrative and boundless, which was different from the pituitary adenoma. The tumor was partially excised. Nevertheless, the patient’s symptoms continued to deteriorate. After another month, the patient was in total coma. By then, MRI delineated an apparent extension of the tumor with massive edema in the right frontal lobe and multiple hemorrhagic lesions (Fig. 3). Three months later, the patient died.

Histologic examination showed neoplastic cell proliferation against a vascular background. Small round-to-oval cells with hyperchromatic nuclei and indistinct cytoplasm were observed, which appeared as neoplastic cell proliferation against a vascular background (hematoxylin-eosin, ×400).

3. Discussion

Sarcomas are relatively uncommon tumors, accounting for 1% of all malignancies. Histologically, more than 30 subtypes have been described [16]. Rhabdomyosarcoma accounts for 19% of all sarcomas, and it is the most common soft tissue sarcoma in children [2]. It is commonly found in the head and neck region, such as orbit [6,8], nasopharynx [9,12,14], temporal bone [4], and sinonasal tract [5]. However, very few cases of primary intracranial rhabdomyosarcoma have been reported in the literature [13,19]. To our knowledge, only 1 case of rhabdomyosarcoma originating primarily within the sella turcica has been reported [3]. Although Jalalah et al [7] had reported a case of rhabdomyosarcoma involving the region of the sellar turcica in 1987, the origin was concluded to be the sphenoid and ethmoid sinus. In the present case, the tumor was
confirmed to have originated from the lower right portion of the sella turcica. The origin of the tumor may have been a store of primitive mesenchyma lodged either in the dura mater of the sellar floor or in the pericapillary space of the pituitary gland.

Rhabdomyosarcomas are immunoreactive to desmin and myoglobin. Reactivity to muscle-specific actin occurs in more than 75% of cases [18]. Because of the glycogen and myofibrils contained, those tumor cells stain positively with the periodic acid-Schiff reagent and phosphotungstic acid [11]. It expresses a DNA binding protein, MyoD1, which may turn out to be a lineage marker for rhabdomyosarcomas [10,15].

Our findings suggest that rhabdomyosarcoma should be considered in the differential diagnosis of a primary intrasellar neoplasm. The treatment of rhabdomyosarcoma is substantially different from that of pituitary adenoma. In the present case, had a correct diagnosis been initially established, a multimodality therapy with combination of chemotherapy and external beam radiation in addition to the surgery might have been considered promptly, which could have been helpful in prolonging the patient’s life.

References

This article documents a rare case of rhabdomyosarcoma arising from the sella turcica. Aggressive characteristics of the tumor are well documented and pathological examination of the tumor tissue confirms the diagnosis of rhabdomyosarcoma. Radiologic characteristics are protein and a preoperative diagnosis is nonspecific.

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