Primary Osteogenic Sarcoma Involving Sella-sphenoid Sinus

—Case Report—

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Abstract

A 38-year-old male presented with an extremely rare primary osteogenic sarcoma, unassociated with Paget's disease or late effects of radiation, involving the sella and sphenoid sinus region. Complete excision of the tumor was achieved through an extended frontobasal approach. Postoperatively, six cycles of combination chemotherapy (adriamycin, ifosphamide, and cisplatin) followed by a total of 55 Gy local radiotherapy in 33 fractions was given. Primary osteogenic sarcoma should be considered in the differential diagnosis of the central skull base tumors. Osteogenic sarcoma, in general, has a bad prognosis, and should be managed aggressively with multimodality treatment including gross total surgical resection, combination chemotherapy, and radiotherapy.

Key words: osteogenic sarcoma, sella-sphenoid sinus mass, skull base tumor

Introduction

Osteogenic sarcoma is the most common primary malignant tumor of the bone after multiple myelomas. Primary osteogenic sarcoma is very rare in the skull bones and even rarer in the central skull base, i.e., sella, sphenoid sinus, and parasellar region. Only two cases of primary osteogenic sarcoma of the sella and sphenoid sinus region have been reported.^{10,14)} We describe another case of primary osteogenic sarcoma involving the sella and sphenoid sinus region.

Case Report

A 38-year-old male presented with a history of episodic diplopia with horizontal separation of images persisting for 5 years. Diplopia was greater on looking to the right. He was treated with steroids at the referring hospital. He had also a history of weight gain. There was no history of erectile impotence or loss of libido or of Paget's disease of the bones. He had not received radiation therapy in the

past.

General physical examination revealed no abnormality. Secondary sexual characteristics were well developed. He was fully conscious and alert. Visual acuity in the right eye was 6/36 and 6/9 in the left eye. Visual fields and fundi were normal. Other neurological findings were unremarkable except for mild right abducens nerve paresis.

Routine hematological and biochemical parameters were within normal limits. Serum alkaline phosphatase level was normal. Relevant endocrinological tests showed that serum T3 level was 120 ng/ml (normal range 100–190 ng/ml), serum T4 was 7.2 µg/dl (5.5–11.5 µg/dl), thyroid-stimulating hormone level was 5.5 µIU/ml (0–10 µIU/ml), serum cortisol was 21.8 µg/dl (9–25 µg/dl), and testosterone was 556 ng/dl (300–1200 ng/ml). Skull radiography showed gross destruction of the sella and upper clivus with calcification. Chest radiography and long bone radiography revealed no abnormalities. Computed tomography (CT) showed an isodense sella-sphenoid sinus mass with areas of calcification, enhanced by contrast medium (Fig. 1 left). Bone views revealed destruction of the sella and upper clivus (Fig. 1 right). Magnetic resonance (MR) imaging revealed a well-defined, lobulated, midline mass le-

Fig. 1 left: Computed tomography (CT) scan with contrast medium showing an enhanced mass in the sella-sphenoid sinus region. right: Bone CT scan showing destruction of the sella and upper clivus.

Fig. 3 Diagram of the operative approach. S: sella, Sp: sphenoid sinus, arrow: approach, hatched area: osteotomy, shaded area: craniotomy.

Fig. 2 A: T₁-weighted magnetic resonance (MR) image, coronal view, showing an isointense mass in the sella and sphenoid sinus. B: T₂-weighted MR image, coronal view, showing a hypointense mass. C: MR image with gadolinium, coronal view, showing a highly enhanced mass in the sella and sphenoid sinus. D: MR image with gadolinium, sagittal view, showing multiple hypointense areas within the mass indicating calcification.

sion in the central skull base, i.e., the sphenoid sinus, sella, clivus, and suprasellar region. The lesion was isointense on the T₁-weighted images (Fig. 2A) and hypointense on the T₂-weighted images (Fig.

Fig. 4 Postoperative computed tomography scan with contrast medium showing the tumor cavity filled with fat graft (arrow).

2B), and enhanced by contrast medium (Fig. 2C, D). The preoperative diagnosis was clival chordoma.

The tumor was totally excised through an extended frontobasal approach on April 7, 1997. The extended frontobasal approach is a modification of the transbasal approach of Derome, with the addition of a orbito-fronto-ethmoidal osteotomy. The osteotomies minimize brain retraction. The tumor was approached extradurally after drilling the planum sphenoidale and ethmoids (Fig. 3). The tumor was grayish white, firm to hard with areas of dense calcification, and was relatively avascular. Destruction of the sella and clivus was evident. The tumor involved the medial walls of both cavernous sinuses. The tumor cavity was packed with autologous fat

Fig. 5 Photomicrographs showing the cellular tumor includes sheets of pleomorphic and hyperchromatic oval to polygonal cells and irregular masses of osteoid (A-D). There is focal calcification (B) within the osteoid. Note the occasional mitosis (C, arrow) and group of osteoclastic giant cells (D). HE stain, $\times 240$ (A, B); $\times 385$ (C, D).

graft. The anterior cranial fossa floor was repaired with fascia lata and vascularized pericranial graft.

The postoperative period was uneventful. Postoperative CT revealed complete excision of the tumor (Fig. 4). The excised specimen consisted of multiple pieces of gray-white, firm to hard tissue, with a total diameter of 4 cm. Histological examination showed the tumor was cellular with sheets of oval to polygonal cells having considerable nuclear pleomorphism and an occasional mitotic figure. Irregular masses of malignant osteoid cells with focal calcification were also present, as well as groups of osteoclastic giant cells (Fig. 5). The diagnosis was osteogenic sarcoma of the sellar region.

The patient was discharged on the 10th postoperative day. Neurologically, he was intact except mild right abducens nerve paresis. He was referred to a cancer center for adjuvant therapy. Six cycles of combination chemotherapy, consisting of adriamycin 40 mg/m^2 on day 1, ifosphamide $1.3 \text{ g/m}^2/\text{day}$ for 3 days, and cisplatin 100 mg/m^2 in three doses on days 1–3, were given. This was followed by a local boost of radiotherapy. A total dose of 55 Gy in 33 fractions were administered over a period of 6 weeks. At follow up, 18 months after surgery, he was

doing well. He was completely recovered of right sixth cranial nerve paresis.

Discussion

Osteogenic sarcoma is a malignant neoplasm primarily occurring in long bones, with the greatest predilection for the metaphyses, most frequently the distal femur and proximal tibia. Osteogenic sarcoma can develop *de novo* or secondary to Paget's disease, fibrous dysplasia, retinoblastoma, chronic osteomyelitis, and after exposure to radiation therapy.⁵⁾ This tumor affects mainly children and young adults with the peak incidence in the second decade, and occurs most frequently in males (60%).⁹⁾ These tumors grow rapidly; the natural history is generally short, with pain and local swelling as the most common symptom. Metastasis to lungs might occur with the appearance of the first symptom.

The radiographic features of osteogenic sarcoma reflect the bone formation, destruction, and periosteal reaction.⁵⁾ Bone spicules radiate from the advancing margins of the osteolytic process, producing a "sunray" appearance; this typical feature is less frequently seen in the skull than in long

bones. In the skull, the spicules of bone are usually associated with thickening of the calvaria at the advancing edge of the tumor, due to subperiosteal reaction.¹²⁾ CT and radiography are superior to MR imaging for detecting the matrix calcification and bone destruction or reaction.⁵⁾ Histological diagnosis of osteogenic sarcoma depends on the identification of malignant spindle cell stroma which directly produces osteoid or immature bone. There are three subtypes based on the predominant cell type forming the lesion: Fibroblastic, chondroblastic, and osteoblastic. In most cases, spindle cell malignant stroma is seen that may imitate fibrosarcoma, but production of osteoid directly by the tumor cells is pathognomonic.⁵⁾

Overall, only 5–10% of osteogenic sarcoma occurs in the craniofacial bones, and most of these are located in the mandible and maxilla. Less than 1% are found in the calvaria,⁵⁾ although 8.6% of osteosarcoma affected the craniofacial bones in one series.³⁾ Primary or secondary occurrence of osteogenic sarcoma is very rare in the skull vault.^{1,4,6–8,11,18–20)} Primary intracerebral osteosarcoma has also been reported.^{9,15)}

Primary osteogenic sarcomas of central skull base, i.e., clivus, sella, parasellar region, are extremely rare with only two cases reported previously.^{10,14)} One was primary osteogenic sarcoma of the sellar region extending forward from the clivus as far as the frontal paranasal sinus occurred in a 49-year-old female, who presented with headache for 1.5 years, and later developed anesthesia over the left side of her forehead and face and left abducens nerve palsy.¹⁰⁾ She died 6 days postoperatively. Another case was of primary osteogenic sarcoma of the sella and sphenoid sinus region occurred in a 22-year-old male who suffered left frontal headache and diplopia.¹⁴⁾ Neurologically, he had bilateral abducens nerve palsies. Biopsy of the lesion was performed through the transnasal transsphenoidal route. Postoperatively, he was treated with radiotherapy and combination chemotherapy.

Our patient also presented with episodic diplopia with mild right abducens nerve paresis. He did not have facial hypesthesia as found in previous cases. Only biopsy of the lesion was performed in the earlier cases, whereas we could achieve total excision of the tumor via an extended frontobasal approach in our case. A similar case of intraosseous, well-differentiated osteosarcoma of the clivus had an indolent course and better prognosis.¹³⁾ Osteogenic sarcoma, in general, has a poor prognosis. This is particularly so with tumors in this location where the diagnosis is usually late and intracranial extension prevents total resection.²⁾ Multimodality

treatment including gross total surgical resection, combination chemotherapy, and radiotherapy is the best approach as shown in our case.¹⁷⁾ In a recent series, the overall survival with radical surgery and multidrug chemotherapy for long bone osteosarcoma has been reported to be 65% at 3 years and 55% at 5 years. The progression-free survival at 5 years was 44%.¹⁶⁾ However, the superiority of combined modality therapy has not been established by controlled clinical trials, but these improved results should serve as a model for more effective treatment of tumors arising in the cranial bones, especially as modern surgical techniques allow wide resection of tumors involving not only the calvaria but also the base of skull.³⁾

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