

Case Report

Ou Young Kwon, M.D.

Sang Koo Lee, M.D.

Maeng Ki Cho, M.D.

Young-Joon Kim, M.D.

Department of Neurosurgery
Dankook University
College of Medicine
Cheonan, Korea

A Case of Biphasic Synovial Sarcoma of Frontal Bone in an Elderly Patient

Synovial sarcomas are rare soft tissue malignancies arising from tendons, tendon sheaths, and bursal structures. These tumors usually develop in the extremities of adolescents and young adults. Uncommonly, these tumors may arise in the head and neck approximately 9% of all synovial sarcomas. Most common sites of head and neck synovial sarcomas are hypopharynx and surrounding structures of paranasal sinuses. However, frontal bone without involving paranasal sinus is extremely rare. We report a case of biphasic synovial sarcoma of the frontal bone discuss the clinical and pathologic features of this case with the literature review.

KEY WORDS : Synovial sarcoma · Paranasal sinuses · Frontal bone.

INTRODUCTION

Synovial sarcomas are aggressive malignant soft-tissue tumors comprising 5-10% of all soft tissue sarcomas. These tumors typically occur in adolescents and young adults 15 to 40 years of age. More than 85% of synovial sarcomas arise in the extremities, mainly in the lower limbs around the knee^{2,3,5,6,11}. Synovial sarcomas are frequently related to tendon, tendon sheaths, and bursal structures. They can arise in joint cavities and may arise anywhere unassociated with a joint cavities including head and neck, lower back, chest and abdominal wall^{5,6,9}. Synovial sarcomas do not always originate from the synovium but instead have a tendency to arise from unknown parasynovial tissues as parapharyngeal, paranasal, retroperitoneal and mediastinal regions^{4,11}.

Among synovial sarcomas in the head and neck region, most of them have been located in hypopharynx³. Few cases have been reported around the paranasal sinuses such as maxillary, ethmoid, frontal and sphenoid sinuses^{1,2,5,8,10,12,13}. We report an unusual case of synovial sarcoma at the frontal bone which is not associated with either synovium or paranasal sinuses in elderly woman.

CASE REPORT

History and examination

The 75-year-old woman presented with slowly growing soft mass on frontal area for three years. She did not have any medication or head trauma history except for operation and radiation therapy on thyroid mass 40 years before admission. Neurologic examination revealed no abnormal findings. Magnetic resonance (MR) imaging demonstrated homogeneously well-enhancing huge mass which was mainly involving on the frontal bone and preserving the frontal sinus (Fig. 1). There were little mass effect on the anterior aspect of the frontal bone. Three-dimensional head computed tomography (3-D head CT) scan revealed extensive destructive skull changes at the frontal

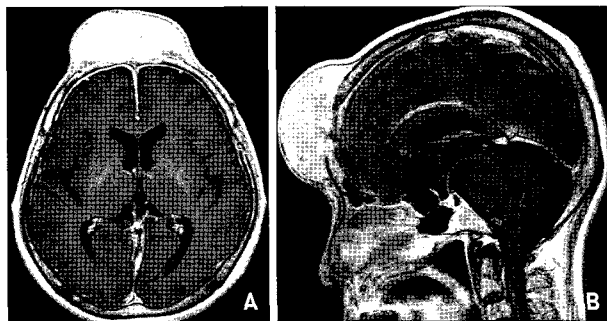


Fig. 1. Preoperative magnetic resonance imaging. Axial (A) and sagittal (B) gadolinium-enhanced T1-weighted images demonstrating a homogeneously well enhancing mass on the frontal bone without involving frontal sinus.

- Received : May 2, 2007
- Accepted : July 1, 2007
- Address for reprints :
Sang Koo Lee, M.D.
Department of Neurosurgery
Dankook University
College of Medicine
16-5 Anseo-dong
Cheonan 330-715, Korea
Tel : +82-41-550-3090
Fax : +82-41-552-6870
E-mail : lsk9999@dreamwiz.com

bone (Fig. 2). There was no destruction of inner table of the skull or frontal sinuses. For the evaluation of the possibility of metastasis or other systemic sarcomas, we also performed metastatic work up including tumor markers, hormone assays, chest and 4-extremities radiographs, abdominal ultra-sonography (US). We did not find any abnormal findings on metastatic work up.

Operative findings

The mass was gross totally removed including involved frontal bone. On operative findings, the mass was fixed to thickened galea aponeurotica and was tightly adhered to surrounding frontal bone. The tumor was 8×7 cm-sized, gray-reddish colored, moderately hard mass. Extensive calcification in the some part of the mass was noticed. After the extracranial mass removal, there were multiple spicule-like changes on the frontal bone. We performed extended craniectomy to remove involved frontal bone. At that time, the frontal sinus was spared and the dura was intact. At the end, the canioplasty with artificial bone was done in the craniectomy site.

Histopathological features

The tumor had biphasic natures with mesenchymal and epithelial components. The specimen showed two populations; one with solid tumor nests having oval round nuclei with

atypia and pale eosinophilic cytoplasm, and the other with gland-forming tumor cells. Immunohistochemically, solid tumor nests were positive for vimentin and gland-forming cells were positive for cytokeratin (Fig. 3). Some portion of tumors were partially reactive for NSE and CD56 but, immunoreactivity for CEA, synaptophysin, chromogranin were not evident. The morphological and immunohistochemical study were characteristic biphasic synovial sarcoma.

Postoperative course

Postoperatively, her clinical course was uneventful. After confirming the tissue diagnosis, we performed adjuvant radiotherapy was scheduled. Intensity-modulated radiotherapy was conducted using 31 fractions and a single iso-center to a total dose of 6200 cGy. Brain MR imaging after 3 months following completion of radiotherapy demonstrated no definite local recurrence. The patient has not shown any neurological deterioration up to present time.

DISCUSSION

Synovial sarcomas are malignant soft tissue tumors closely related to tendons, tendon sheaths, and bursal structures⁵. In spite of their name, these tumors do not always arise from synovial tissues. They are so named because of their histologic similarity to synovium. So, they can arise from any portion of the body containing pluripotent mesenchymal cells near or even remote from articular surfaces, tendons, tendon sheaths, juxta-articular membranes and facial aponeuroses^{6,10,11}. Uncommonly, these tumors may arise in the head and neck approximately 9% of all synovial sarcomas. In the head and neck region, hypopharynx is the most common involving site probably due to having abundant synovial tissues¹⁰. But, synovial sarcomas involving paranasal sinuses have rarely been reported. The first description of paranasal sinus synovial sarcoma was reported in 1970 by Tribble¹³ who described a case of metastasized synovial sarcoma to the sphenoid sinus. Since that time, there have been only 7

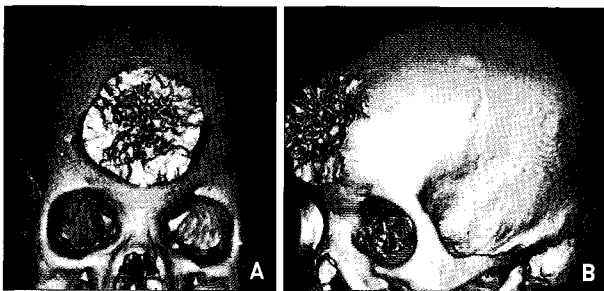


Fig. 2. Preoperative 3D head computed tomography scans (A and B) demonstrating the destructive changes of the frontal bone. The mass is not penetrating into inner table of skull, but deforming outer table to creator-like pattern.

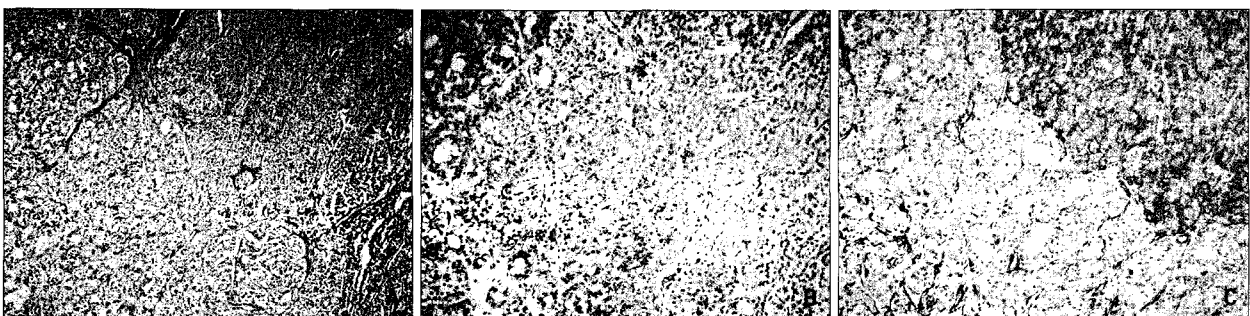


Fig. 3. Histologic findings of biphasic synovial sarcoma. Microphotograph of solid portion shows classic biphasic synovial sarcoma with glandular structures surrounded by a spindle cell component (A : H&E stain, × 200). Immunohistochemistry for cytokeratin highlights the epithelial elements (B : cytokeratin[AE1/AE3], ABC method, × 200). The spindle cell elements are stained brown for vimentin stain (C : vimentin, × 200).

Table 1. Summary of cases of synovial sarcoma involving paranasal sinuses

Authors(year)	Age/Sex	Sinus Location	Treatment	Histology	Outcome / FU period
Trible ¹³ (1970)	24/F	sphenoid	Op+RTx	NR	died / 2 yrs
Chiak et al. ² (1997)	25/M	maxillary	Op+chemo+RTx	monophasic	recur/ 2 yrs
Bangheard ⁵ (2001)	12/M	maxillary	Op+chemo+RTx	NR	NED / 2 yrs
Kartha et al. ⁸ (2002)	24/F	ethmoid	Op+RTx+chemo	monophasic	died / 9 mos
Sun et al. ¹² (2003)	54/M	maxillary	Bx+RTx+Op	biphasic	NED / 45mos
Bettio et al. ¹ (2004)	36/M	sphenoid	Op	monophasic	died / 1 wk
Gary et al. ⁵ (2005)	44/M	frontal	Op+RTx+chemo	monophasic	NED / 2 mos

*Abbreviations: chemo: chemotherapy, FU: follow up, RTx: radiation therapy, NED: no evidence of disease, NR: not reported

reported cases of synovial sarcomas involving the paranasal sinuses (Table 1)^{1,2,5,8,10,12,13}. Patients were male predominant and ranged in age from 12 to 54 years (mean 31 years). But, our case was unusually old-aged patient whose tumor was located in the frontal bone without involving frontal sinus. We assumed that this case have the possibility of sarcomatous change from galea aponeurotica. On operative findings, there were thickened galeal layers around the tumor base. There might have some relationship between previous radiation for thyroid mass 40 years ago and unusual synovial sarcoma.

Histologically, synovial sarcoma is a mixture of epithelial and spindle cell components, occasionally exhibits a biphasic patterns. If this tumor shows only one cell type, it can be classified as monophasic spindle cell type or monophasic epithelial cell type synovial sarcoma^{3,5,8,10}. Jernstrom⁷ defined the synovial sarcoma as a biphasic tumor that is "histologically composed of two sharply contrasted types of tissues; one type reproduces caricatures of synovial structure, the other consist of fibromatous element. Therefore, biphasic synovial sarcoma can be easily diagnosed by characteristic histopathologic findings. But, monophasic variants exist which can be difficult to diagnose. Synovial sarcomas are characterized by a specific balanced translocation (X; 18), which is found in greater than 90% of all synovial sarcoma subtype. This translocation has not been found in other sarcomas or carcinomas^{2,10}. This chromosomal study can be helpful when histological studies are equivocal. Cytogenetic analysis and molecular techniques to detect translocation have aided in diagnosing synovial sarcomas, especially monophasic fibrous types and poorly differentiated forms that can be difficult to differentiate from other types of sarcomas^{2,5}. Rhabdomyosarcoma, liposarcoma, leiomyosarcoma, angiosarcoma, minor salivary gland pleomorphic adenomas, osteosarcoma should be included in differential diagnosis^{6,8}.

Treatment guideline is limited because there are small cases in paranasal sinus region (Table 1), so it is based on experiences with tumors developing in the extremities. Primary therapy in most patients with synovial sarcomas is operative excision followed by adjuvant radiotherapy or chemotherapy⁷. Some authors advocated that postoperative radiation therapy improved

local control rates and chemotherapy may prevent or delay the occurrence of metastasis¹⁰. But, until now there have been debates concerning the role of radiation therapy or chemotherapy for synovial sarcomas of both extremities and head and neck. Postoperative radiation therapy for synovial sarcomas has not been found to increase survival rate, although it has improved local

control rates⁸. Moreover, it seems to be difficult to determine how beneficial chemotherapy is as the treatment modalities of synovial sarcomas. In present case, the patient underwent surgical excision first and followed by adjuvant radiotherapy according to recommendation.

The reported 5-year survival rates of patients suffering synovial sarcomas were from 36 to 76% ; the 10-year survival rates were from 20 to 63%¹⁴. Some authors advocated that patient's prognosis is closely related to histological findings depending on well-differentiated or poorly-differentiated sarcomas⁹. Although synovial sarcomas developing in the extremities show similar histological findings in other regions, it is usually thought that head and neck synovial sarcomas have the better prognosis^{6,8}. This may be because synovial sarcomas in the head and neck present in relatively younger patients and are treated earlier after the onset of symptoms^{6,12}. Duvall and associates³ analyzed that the size of the tumor was another prognostic factor. Tumor size less than 4 cm in diameter showed better prognosis than those with larger tumors. On the other hand, Kartha⁸ emphasized that outcome was not related with the extent of tumor, but timing of radiation and local recurrence. In our case, frontal synovial sarcoma was bigger size than previous reports. Thus, regular follow up evaluation and close observation will be needed for possibility of local recurrence.

CONCLUSION

Synovial sarcomas can arise from the frontal bone without involving paranasal sinus. Nevertheless, it should be considered that there is possibility of unusual type of sarcomas, to be included in the differential diagnosis, of frontal masses. Despite its rare incidence, its clinical behavior, optimal treatment and prognosis of synovial sarcomas should be appreciated.

References

1. Bettio D, Rizzi N, Colombo P, Bianchi P, Gaetani P : Unusual cytogenetic findings in a synovial sarcoma arising paranasal sinuses. *Cancer Genet Cytogenet* 155 : 79-81, 2004
2. Chiak RA, Lyndiatt WM, Lyndiatt DD, Bridge JA : Synovial sarcoma of the head and neck : chromosomal translocation (X;18) as a diagnostic

- aid. *Head Neck* 19 : 549-553, 1997
3. Duvall E, Small M, Al-Muhanna AH, Maran AD : Synovial sarcoma of the hypopharynx. *J Laryngol Otol* 101 : 1203-1208, 1987
 4. Fisher C, Montgomery E, Healy V : Calponin and h-caldesmon expression in synovial sarcoma ; the use of calponin in diagnosis. *Histopathology* 42 : 588-593, 2003
 5. Gallia GL, Sciubba DM, Hann CL, Raman SP, Westra WH, Tufaro AP, et al : Synovial sarcoma of the frontal sinus. *J Neurosurg* 103 : 1077-1080, 2005
 6. Hirsch RJ, Yousem DM, Loevner LA : Synovial sarcomas of the head and neck : MR findings. *AJR Am J Roentgenol* 169 : 1185-1188, 1997
 7. Jernstrom P : Synovial sarcoma of the pharynx ; Report of a case. *J Clin Pathol* 24 : 957-961, 1954
 8. Kartha SS, Bumpous JM : Synovial cell sarcoma : diagnosis, treatment, and outcomes. *Laryngoscope* 112 : 1979-1982, 2002
 9. Morton MJ, Bergquist TH, McLeod RA, Unni KK, Sim FH : MR imaging of synovial sarcoma. *AJR* 156 : 337-340, 1991
 10. Rangheard AS, Vanel D, Viala J, Schwaab G, Casiraghi O, Sigal R : Synovial sarcomas of the head and neck : CT and MR imaging findings of eight patients. *AJNR Am J Neuroradiol* 22 : 851-857, 2001
 11. Sigal R, Chancelier MD, Luboinski B : Synovial sarcomas of the head and neck : CT and MR findings. *AJNR Am J Neuroradiol* 13 : 1459-1462, 1992
 12. Sun JJ, Rasgon BM, Wild TW, Hilsinger RL Jr : Synovial cell sarcoma of the maxillary sinus : a first reported case. *Otolaryngol Head Neck Surg* 129 : 587-590, 2003
 13. Tribble WM : Destructive lesions of the sphenoid. *South Med J* 63 : 849-852, 1970
 14. Weiss SW, Goldblum JR : *Enzinger and Weiss's Soft Tissue Tumors*, ed(4). St. Louis : Mosby, 2001