PRIMARY EWING'S SARCOMA OF THE ORBIT

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Ewing's sarcoma, first described by James Ewing's(1), accounts for approximately 10% of primary malignant bone tumors(2). These tumors primarily occur in the long bones, pelvis, and ribs in 47, 29, and 12% cases respectively(3). The skull is primarily involved in 1-6% of all cases and these usually occur in the frontal and parietal convexities(4-6). Ewing's sarcoma primarily arising in the base of skull is distinctly rare(5). Cases originating from the ethmoid(7) and intra-cranial portion of the roof of the orbit(8) have been reported. We report a case of localized primary Ewing's sarcoma of orbit originating from the lesser wing of sphenoid and presenting clinically as a painful proptosis.

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Received for publication: July 15, 1992; Accepted: March 15, 1993

Case Report

A 10-year-old girl was admitted with complaints of painful progressive prominence of the left eye ball for 2 months and failing vision in the left eye for 1 month. Examination revealed a non pulsatile, non-reducible proptosis, chemosis, congestion and medial deviation of the left eye ball. Eye movements were restricted in lateral and superior directions with diplopia. The left visual field was constricted. There was no other clinical abnormality. Hematological investigations were normal. Plain roentgenogram showed destruction of the lateral wall (lesser wing of sphenoid) of orbit. CT scan showed a mixed attenuation mass in superior and lateral portions of the left orbit and apex of temporal fossa contiguous with an expanded and eroded lesser wing of sphenoid. The tumor showed uneven contrast enhancement with patchy hypodense areas within the lesion (*Fig. 1*). A complete skeletal survey for associated lesions, abdominal CT and skiagram of chest revealed no abnormality.

A left fronto-temporal craniotomy and orbitotomy was performed. Necrotic material mixed with old clots gushed out as soon as the orbital wall was removed. This moderately vascular tumor occupied the superolateral and posterior portions of the orbit and apex of the temporal fossa extradurally. The periorbital and temporalis muscle were infiltrated. A subtotal excision was performed. Histopathological examination of multiple small fragments of tumor showed a monomorphic round cell tumor. Individual cells had scanty cytoplasm and large nucleus with fine peppery chromatin. The tumor cells were supported by very delicate thin vascular stroma, at places having perivascular arrangement of tumor cells (*Fig. 2*). Focal cytoplasmic glycogen was demonstrated in

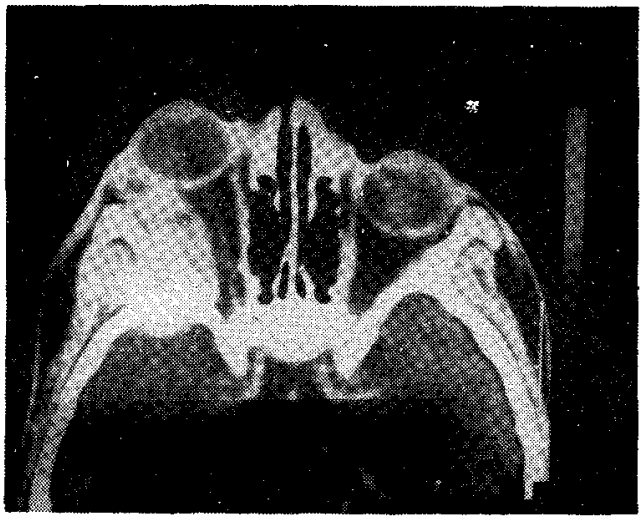

Fig. 1. Contrast enhanced CT scan showing expansion and erosion of both tables of the lesser wing of sphenoid on the left side, with contiguous hyperdense mass (with hypodense areas within it) in the orbit and temporal fossa.

some of the tumor cells on PAS stain. Necrosis and mitosis was evident. In view of the age of the patient, location of the tumor in bone with extension into soft tissues and round cell morphology, a diagnosis of Ewing's sarcoma was made. 40 Gy irridiation was delivered in 15 fractions over three weeks. She received vincristine 1 mg and cyclophosphamide 400 mg intravenously at weekly intervals for 12 weeks. Adriamycin 20 mg intravenously was added to 3 courses. Follow up 1½ years after surgery, revealed no clinical or radiological evidence of residual or recurrent tumor, or any metastasis.

Discussion

Ewing's sarcoma is primarily a rare tumor of the pediatric age group, with 75% occurring below the age of 20 years. Its peak incidence is between 5-13 years(6,9). There are only few reports of Ewing's sarcoma originating primarily in the base of skull from petrous(10,11), mastoid(12), ethmoid bones(7), and the intracranial portion of the orbital roof(8). The tumor in the present case arose from the lesser wing of sphenoid as demonstrated by CT.

Ophthalmoplegia, proptosis and blindness due to secondary involvement of the skull by Ewing's sarcoma have been described earlier(13) but a primary intraorbital location of the tumor has not been reported. The common symptoms of pain and swelling with peripheral location of Ewing's sarcoma(14) explain the painful proptosis in the present case. Congestion, chemosis and restriction of eye movements were probably the result of compression or direct invasion by the extradural tumor(15).

The CT morphology of diffuse enhancing mass lesion, described in 3 cases of primary Ewing's sarcoma of the skull was also observed in the present case(8,11,16). Expansion and destruction of the lesser wing of sphenoid with loss of cortical bones in both tables, and contiguous location of the tumor external to temporal dura and periorbita suggested its origin from the lesser wing of sphenoid. The hypodense areas observed on CT represented old hemorrhage and necrosis within the tumor.

The postulated myelgenous multifocal origin with a manner of spread similar to plasma cell myeloma, supports the basis for subclinical metastasis in this tumor(17). They commonly occur in the lung and bone in 75-85% cases during the first 2 years(18,19). However, the clinical examination and roentgenographic skeletal and chest survey before and one year after surgery were normal in the present case. Histopathologically, the possibility of rhabdomyosarcoma was not considered due to this being a round cell tumor primarily arising in the bone, the absence of differentiation of cells into myoblasts and the presence of glycogen focally within the tumor cell cytoplasm.

A 50-80% disease free survival at 2 years with new multimodal protocol, including

Fig. 2. Microphotograph showing uniform cells with scanty cytoplasm and mitotic figures (Hematoxylin and Eosin × 480).

chemotherapy, has been reported(20,22). Both local disease control and prevention of systemic metastases contribute to a better outcome. The peripheral location of tumor and absence of metastases at the time of diagnosis indicate good outcome. Prognosis of Ewing's sarcoma arising from the cranium may be similar to that for the other sites(4-6,16). The follow-up is too short to predict her chances of cure accurately.

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NOTES AND NEWS

FIFTEENTH SCIENTIFIC MEETING OF THE INTERNATIONAL SOCIETY OF HYPERTENSION

The 15th Scientific Meeting of the International Society of Hypertension will be held at Melbourne (Australia) from *March 19-26, 1994.*

For further details please contact: Dr. Shailendra Vajpeyee, President, Indian Society of Hypertension, Govt. Medical College, Surat 395 001, Gujarat. Phone: (Off.) 46130, (Res.) 41371, Fax : 91-261-652338.

A Satellite Symposium of ISH 1994 Melbourne Meeting on Hypertension and Heart Research in Developing Countries will be held in Bombay on *March 14-15, 1994.*

For further details please contact: Dr. K.G. Nair, President, Organizing Committee, 206, Doctor House, Pedder Road, Bombay 400 026, Phone: 3865008, Fax: 91-22-467780.