Solitary Intracranial Plasmacytoma In A Child Report of a 7-year-old girl

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Abstract : A case is reported of large intracranial plasmacytoma arising from the dura mater and involving the dura of the left cerebral convexity, tentorium cerebelli and the dura of the posterior fossa in a 7yearold girl is reported.

Key Words : Childhood neoplasm, Computed tomography, Plasmacytoma, Surgery

INTRODUCTION

Involvement of the central nervous system is one of the commonest complications of multiple myeloma and is most frequently manifested by spinal cord compression secondary to bony infiltration. Numerous cases of intracranial plasmacytomas with a dural attachment have been reported (3,4,11, 12,16,20,23,24,27,29,30,32,34,35,40,). We report a 7yearold girl who showed a huge intracranial spaceoccupying lesion on computerized tomographic (CT) scans. Neuroradiological features, surgical treatment and radiotherapy of this unusual lesion were discussed in the light literature.

CASE REPORT

A 7 year-old girl was admitted in May 1988, with the chief complaint of headache. The occipital headache which began months prior to admission. She had had several episodes of nausea and vomiting prior to admission. Neurological examination was normal except for bilateral chronic papilloedema. An electroencephalogram (EEG) showed focal delta waves in the left occipitoparietal area. CT revealed an extracerebral mass in the left parietooccipital convexity, which was enhanced after the infusion of contrast medium (Fig.1). Because of the patient's reaction during intravenous infusion of iodinated contrast medium, she was not subjected to angiography. Xrays roentgenograms of the skull showed impressio digitata (Fig.2).

Operation: A wide parietooccipital scalp incision wasd made. The outer surface of the parietooccipital bone appeared partially destroyed by tumour. Craniectomy was performed. The under surface of the bone was loosely attached to the tumour in the midportion of the bone, where the inner table was also destroyed and there was an adjacent reactive hyperostosis. The dura mater was thickened, darker and more vascular than normal. The tumour bulged from the dural surface. The dura mater was incised around the entire periphery of the tumour. The tumor was lifted from the compressed cerebral cortex and separated from it cleanly. Although there were adhesions in several areas where pial vessels entered the tumour, total removal accomplished. The tumor had been situated mainly in the subdural space and the gross impression was that arose from the dura mater. The tumour which measured 10 cm in rostrocaudal length and 7x6 cm in the coronal plane consisted of fleshy redbrown tissue.. Cranioplasty was not performed.

A small piece of tissue was embedded in paraffin and sections were stained with hematoxylin and eosin. The histological appearance of the specimen

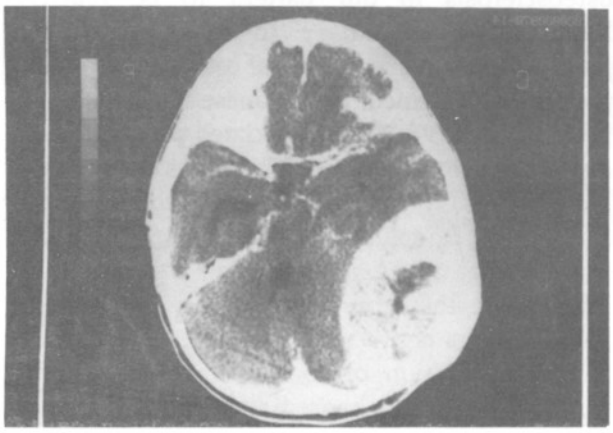

Fig. 1 : Contrast-enchanced axial computed tomographic scan showing an extracerebral mass in the left parieto-occipital convexity.

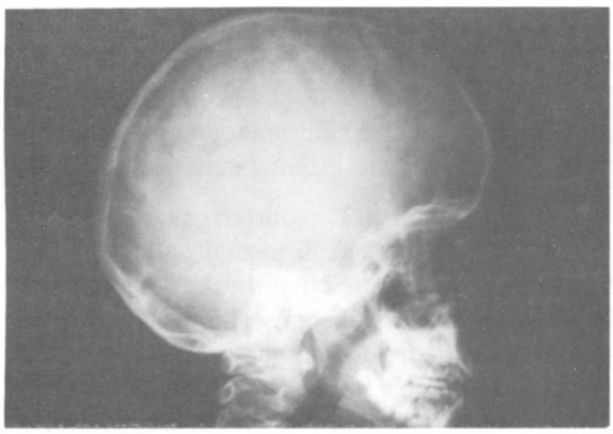

Fig. 2 : Plain x-ray of the skull showing hyperostosis in parieto-occipital area and impressio digitate.

was that of a highly cellular neoplasm composed of mature plasma cells with round, eccentrically nuclei containing coarse clumps of peripheral chromatin and abundant cytoplasm (Fig.3A,3B).

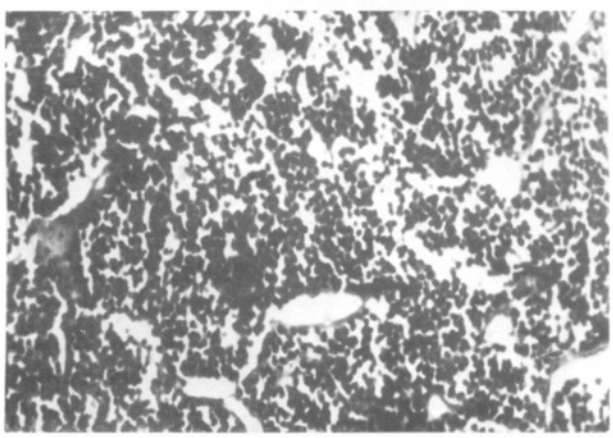

Fig. 3 A : Plasma cells with narrow cytoplasm, round eccentrically placed nuclei which fill the compartments divided by fibrous septa are seen (H&E,x40).

After the diagnosis of plasmacytoma additional laboratory examinations were performed. Complete blood count was within normal limits. There were 8.2mg of serum calcium, 5.1mg of phosphorus per dl. A spinal puncture 6 days after the operation revealed clear spinal fluid containing a small number mononucleocytes and a total protein of 80mg/dl. No further detailed cytology of the CSF cells was performed. Serum protein electrophoretic pattern, albumin 54.3% (Normal 5565%), alpha 1 globulin 5.2% (Normal 15%), alpha 2 globulin 10.4% (Normal 69%), beta globulin 12.9% (Normal 913%), gamma globulin 17.2% (Normal 11-18%). Total serum protein level was 7.8mg/dl. Bence Jones protein test was negative on three occasions. Sternal bone marrow was normal, no evidence of plasma cells in bone marrow was found. Protein electrophoresis showed. IgA 210mg%, IgM 271mg%, IgG 1830mg%. IgM and IgG levels were minimal high.

X-ray bone survey was negative and a bone scan of the entire skeleton showed no abnormality except in the parietooccipital area. The early postoperative period was uneventful except for a collection of cerebrospinal fluid under the skin flap which had to be aspirated.

Radiotherapy was started on the twentieth postoperative day. A dose of 4000 rads in 20 treatments, over five weeks was given to the entire head.

Computed tomography of the brain performed seven months after the operation revealed a collection of cerebrospinal fluid under the skin flap and no tumour recurrence (Fig. 4). Eight months after the operation, a medium pressure ventriculoperitoneal

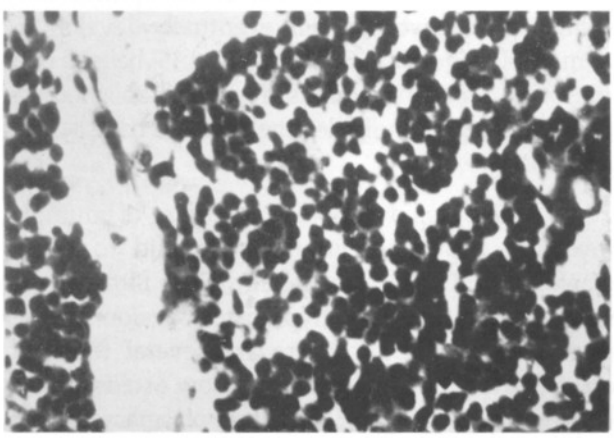

Fig. 3 B : A haematoxylin-eosin-stained photomicrograph showing the presence of a celluler population consisting of plasma cells (x100).

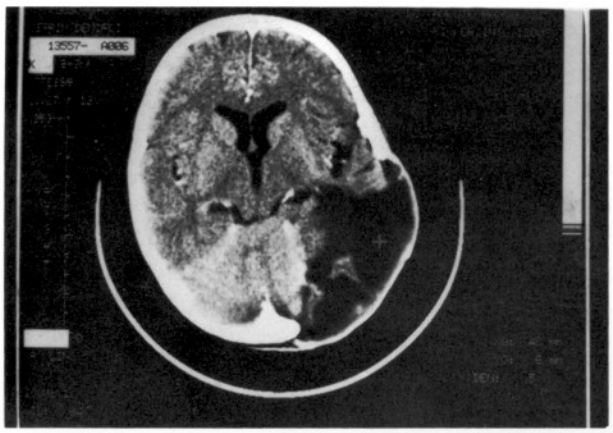

Fig. 4 : Postoperative axial Ct scan showing a collection of cerebrospinal fluid under the skin flap and no tumour recurrence.

shunt device was inserted from the CSF collection under the skin flap to the peritoneum.

DISCUSSION

Within the group of solitary craniocerebral plasmacytomas two subsets can be recognized: those originating in the bone and those originating in the dura mater.

Nine cases of solitary plasmacytoma of the skull base (1,5,7,11,15,18,33,37,38), 16 cases of solitary plasmacytoma with extension into the subdural space (3,4,11,16,20,23,24,27,29,30,34,35,40), 14 cases of plasmacytoma arising from calvarium (2,6,9,11,19, 22,25,28,36) and 5 cases of intraparenchymal plasmacytomas (8,13,14,24,41) are reported in the literature. Our case is a solitary intracranial plasmacytoma in a child.

Some authors (11,17,26) are of the opinion that solitary plasmacytomas will eventually develop into multiple myeloma. Others (9,10,18,35) believe that true solitary plasmacytomas are well established entities unrelated to multiple myeloma and have a good prognosis.

In general, as a radiological feature of multiple myeloma a punchedout lesion is said to be of diagnostic value. In our patient XRay films of the skull demonstrated on hyperostotic lesion on the parietooccipital bone. There are several reported cases of multiple myeloma that show osteosclerotic bone lesions (15,39), but in solitary plasmacytoma of the calvarium, XRay films of the skull demonstrated an osteolytic lesion (1,2,9,19,22,25). We consider the osteosclerosis in our patient to be reactive hyperostosis secondary to invasion of the plasmacytoma. A special type of osteosclerotic multiple myeloma termed, the Japanese Syndrome. has been reported (39). This is associated with endocrine disturbance. In our patient, there was no sign suggestive of the syndrome. Kurtcher and colleagues (25) reported angiographic findings of plasmacytoma. We consider that it was impossible on angiography to differentiate this from meningioma. CT scans of plasmacytoma show a high density mass after intravenous infusion of contrast medium and invariably show an enhanced area (15,21,31). İn our case, CT scan revealed a high density mass after infusion of contrast medium.

Plasmacytoma is a highly radioresponsive tumour. All cases of solitary plasmacytomas of the calvarium reported in the literature have been treated by surgery and radiotheray (2,9,11,19,22,25). Several authors stress that it's sufficient diagnosis and then treat with conventional external radiotherapy (23,38).

Arienta et al (2) reported two cases of plasmacytoma of the skull treated only by radical operation. They stressed that the is good prognosis for patients affected by solitary plasmacytoma of the calvarium who undergo only radical operation, but radiotherapy should be used in cases where surgery does not allow complete removal of the tumoral lesion. Our patient was treated by complete surgical resection followed by radiation therapy. Five years after the operation; there was no neurological deficit in our case.

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