SOLITARY POSTERIOR FOSSA PLASMACYTOMA IN MULTIPLE MYELOMA

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SUMMARY
Neurological complications are well known findings in the course of multiple myeloma disease. They rarely occur as intracranial lesions. There are a restricted number of cases of solitary plasmacytoma or plasmacytoma with multiple myeloma disease in the literature. Infratentorial location of plasmacytoma is extremly rare. In this paper a case of posterior fossa plasmacytoma with multiplemyeloma disease is presented and classification of this pathology, the aid of computerized tomography in the diagnosis and treatment are discussed.

Key words : Plasmacytoma, Multiple myeloma, Posterior Fossa, Cerebral Neoplasm.

Neurological complications are well known findings in the course of multiple myeloma disease. Neurologic complications are frequently in the form of spinal cord or root compression. The pathology, both in the form of isolated lesions or lesions with systemic disease is very rare. They rarely occur with intracranial lesions (1, 2, 3).

The literature contains only a restricted number of cases of solitary plasmacytoma or plasmacytoma with multiple myeloma disease (4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14). Involvement of posterior fossa is extremly rare (15, 16, 13).

The purpose of this paper is to present a case of posterior fossa plasmacytoma with multiplemyeloma; to underline the importance of computerized tomography in diagnosis and to discuss the treatment of this pathology.

CASE REPORT

A 63-year-old male patient admitted to our clinic, complaining of mass lesion at the right occipital region. He was doing well until he noted the onset of headache and a mass lesion in the right occipital region which developed within a two months period. Two weeks ago ataxia and staggering to the right side and a week ago vorniting and nousea had developed.

In neurologic examination, the patient was fully alert, intelligent and cooperative. There was a palpable mass about 4 x 4 cm in size in the right retroauriculer - occipital area. The occipital bone was involved by this hard tissue. Papilledema and cerebellar disturbances were evident.

In serum protein electrophoresis, there were abnormal elevation in beta and gama fractions. Quantative value of immunoglobulin A was found 2100 mg % (normal level : 130 - 430 mg %). Examination of bone marrow obtained by aspiration from sternum revealed an increase in plasma cells.

Routine X rays of skull demonstrated multiple osteolytic lesions of which the right occipital one was the widest (Fig. 1). Computerized tomography revealed a huge, well demarcated hyperdence mass lesion

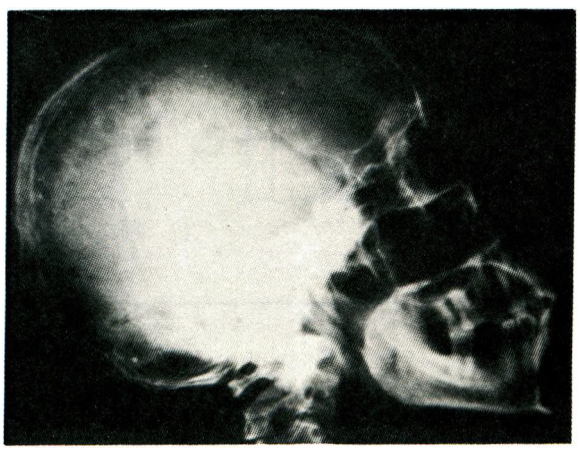

Figure 1. Multipleosteolytic lesions are seen on plain skull radiogram. Destructive area on right occipital bone is evident.

which showed compression on the right cerebellar hemisphere (Fig. 2). The fourth ventricle was deplaced to the left side accompained with obstructive hydrocephalus.

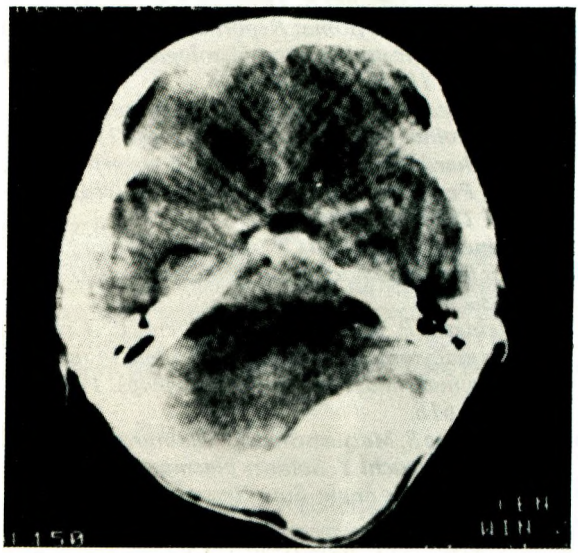

Figure 2. Computerized tomography revealed a huge well demarcated, hyperdence mass lesion compressing on the right cerebellar hemisphere and deplaced fourth ventricle to the left side.

The patient was operated on for harbouring a tumor of posterior fossa. The mass was removed totally. It extended to the bone tissue and was situated extradurally. There was a slight dural infiltration, but no intradural extention. Histopathological examination revealed a plasmacytoma. There was diffuse infiltration of monomorphic plasma cells extending to the bone lamella (Fig. 3). The normal bone marrow was completely destroyed by the tumor cells. The general condition of the patient was well after the operation. Radiation therapy was started immediately in the early post operative period. He received a total dose of 5000 rads, fractionally over 15 days. The follow up examination was performed after 8 months of discharge. Patient's general condition was normal and there was no recurrency of tumor.

DISCUSSION

In the literature there is a confusion about the definition of terms plasmacytoma and multiple myeloma. Although solitary extramedullary plasmacytoma is accepted as a benign lesion according to Dolin and Dewar (17), Carlson et al (18) have pointed out that solitary extramedullary plasmacytoma is an early finding of multiple myeloma disease. Neurological complications frequently occur in multiple myeloma disease. Spinal cord complications appear to be rare. Clarke (4) divided the complication of intracranial involvement into the following three groups. 1. Syndromes of cranial nerve palsies 2. Intracranial tumor syndromes 3. Intraorbital tumor syndromes. Intracranial tumor syndromes is also divided into two groups. The first group consists of mass lesions arising at the lytic area of the bone extending into the cranial cavity. The dura may or may not contribute to the lesion. It frequently constitutes a barrier to the tumor tissue (4, 7, 12). The second group is made of isolated mass lesions, that may develop during the course of multiple myeloma disease (4, 19, 16). No lytic areas can be seen on the skull radiograms of these cases. In this classification only multiple myeloma disease is considered. But solitary plasmacytoma which can appear as an early manifestation of multiple myeloma disease is not mentioned. We think that a third subgroup which can explain the isolated mass lesion developing without any evidence of multiple myeloma must be added to this classification (5, 7, 8, 9, 10, 11, 12, 13, 14). Our literature review showed that most of the lesions were situated in the supratentorial region. Infratentorial localization was found only in a few cases (15, 16, 14). Our case is an example for this rare localization. Another remarkable point of our patient is his admittance with complaints of a cranial mass lesion and neuronal impairment but his lacking any complaints due to a previous multiple myeloma disease.

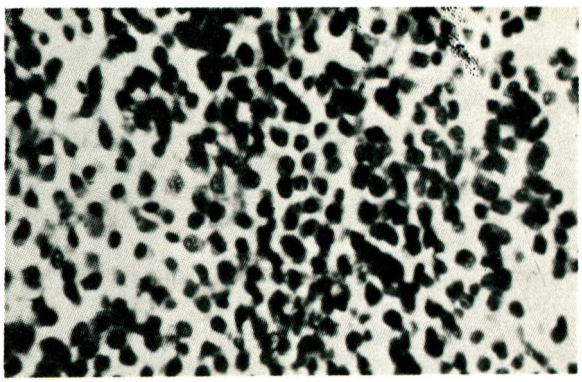

Figure 3. Diffuse infiltration of monomorphic plasma cells was extended to the bone lamella. The normal bone marrow was completely destroyed by the tumor cells. (H.E x 200)

Punch out lesions on routine skull-X rays are the characteristic finding of multiple myeloma disease (20, 21). These punch out lesions can be surrounded by sclerotic areas. In computerized tomography examination, hyperdence, well demarcated lesions can be seen. If the patient has no systemic evidence of multiple myeloma disease than these lesions can be misdiagnosed as a meningioma (7, 8, 16.). There are several cases demonstrating this misdiagnosis. In our case, computerized tomography revealed a well demarcated hyperdence lesion with 38 Hounsfield Unit radioabsorption value. It showed a slight enhancement after intravenous injection of contrast media and eroded the neighbouring cranial bones. We think that these findings are not typical for multiplemyeloma disease or plasmacytoma and especially meningioma, glioma and methastatic tumors

must be considered in the differential diagnosis (7, 8, 16, 12).

Abnormal protein levels were found in serum immunoelectrophoresis. It is also important to determine the levels of quantitative amounts of immunoglobulin in the blood (7, 8, 19, 16, 12). In the present case, immunoglobulin A level was found to be very high while immunoglobulin G and M stayed between the normal levels.

Immunoelectrophoresis of cerebrospinal fluid may be quite helpful for correct diagnosis (15, 8, 9, 14). Especially this laboratory study can be useful in the diagnosis of solitary intracranial plasmacytoma without any evidence of sytemic disease. Bone marrow examination is also helpful for definite diagnosis (22, 16).

Favorable results were obtained by complete or incomplete exicion of tumor with radiation therapy. Some authors believe that radiation therapy alone would be sufficient because intracranial plasmasytoma are listed among radiosensitive tumors (23, 24, 3). However we believe that when the tumor is localized within the posterior fossa, surgical intervention is mandatory.

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