CASE REPORT
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Solitary Plasmacytoma in the Thoracic Spine with Massive Aggregation of Histiocytes

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A Solitary plasmacytoma of bone(SPB) is a rare disease. This paper reports a case of SPB with massive aggregation of histiocytes known as pseudo-Gaucher cells. A 40-year-old male presented with progressive paraparesis and paraesthesia. The Magnetic Resonance Image(MRI) revealed a tumor mass in the thoracic spine that occupied the three colum. The tumor invaded the epidural space of T6 vertebra with compression of the spinal cord. There were no laboratory abnormalities. It was gross totally resected and his neurological symptoms improved. The microscopic examination revealed a plasmacytoma with massive aggregation of histiocytes. Futher neuroradiological studies were carried out and no other lesions were detected.

KEY WORDS : Plasmacytoma · Pseudo-Gaucher cells · Thoracic spine.

Introduction

Solitary plasmacytoma of bone(SPB) accounts for only 3% of all plasma cell tumors. Most cases involve the thoracic spine and there is a strong male predominance. The average age of the patients is older than 50 years of age. We encountered a case of SPB in the thoracic spine that was successfully treated with a surgical resection and adjuvant radiotherapy[6]. Its histopathological findings were a plasmacytosis with massive aggregation of histiocytes, which is known as pseudo-Gaucher cells[7].

Case Report

A 40-year-old male complained of back pain started one-month before the admission and was given conservative medication. However, his pain remained and progressive paraesthsesia and paraparesis developed during the next month. At that time, he was hospitalized, where a physical examination was carried out. He complained of local back pain in the midthoracic level and both lower extremities at exercise. Neurological examination revealed paraparesis of gread IV-, numbness below T3 sensory dermatome and anesthesia in the T7 sensory dermatome. The laboratory examinations were normal (Fig. 1).

On MRI studies, the mass lesion was iso or slightly hyper

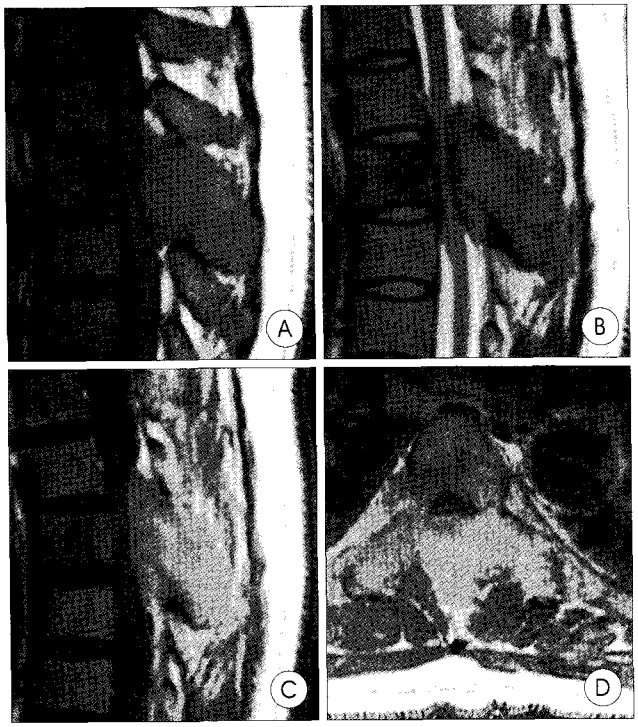

Fig. 1. Sagittal magnetic resonance images of the thoracic spine showing a tumor mass of T6 : iso or slightly hyper intense on T1-weighted image(A), hypointense on T2-weightes image(B) and weak homogenous enhancement(C). The axial image after enhancement(D) shows the tumor mainly invaded the posterior and middle element of T6 compromising the spinal canal.

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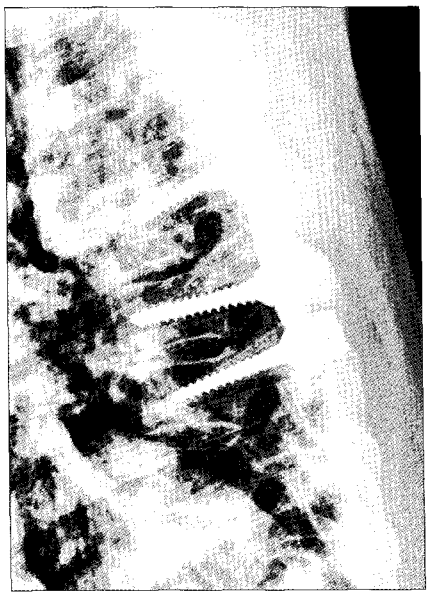

Fig. 2. Postoperative lateral radiograph demonstrates the posterior stabilization from T5 to T7 using the methylmethacryo and the rod and screw system.

intense on T1-weighted image, hypointense on T2-weighted image and showed weak homogenous enhancement. It invaded the spinous process, both transverse processes and pedicles, and posterior half of T6 vertebral body compromising the spinal canal and endangering the spinal cord. Computed Tomograpy (CT) scan was done under the impression of a metastatic spine tumor and no other primary tumors were found. The tumors were resected using a T6 total laminectomy, and both transpedicular approach. The mass was gross totally removed under a surgical microscope. The gel typed whitish color, well capsulated mass had scanty blood vessels and located in the epidural space invading the lamina, pedicles and T6 body. After removing the mass, methylmethacryo was applied into the at corpectomy site and posterior fixation was performed from T5 to T7 using the TYRA rod & screw system (Taeyuon medical. Korea) (Fig. 2).

The microscopic examination of the tumor specimen revealed diffuse proliferation of plasm cells in the bone marrow, which were admixed with histiocytes with abundant eosinophilic granular cytoplasm that is generally known as pseudo-Gaucher cells (Fig. 3). In the other area, there was a massive aggregation

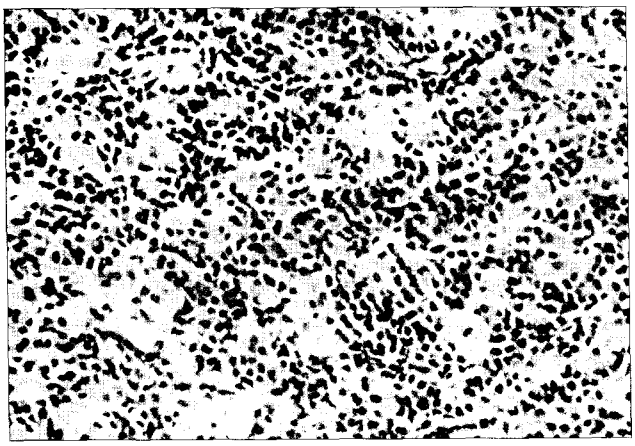

Fig. 3. There is a diffuse proliferation of plasma cells, along with histiocytes that have abundant cytoplasm, so-called pseudo-Gaucher cells. (H&E, ×200).

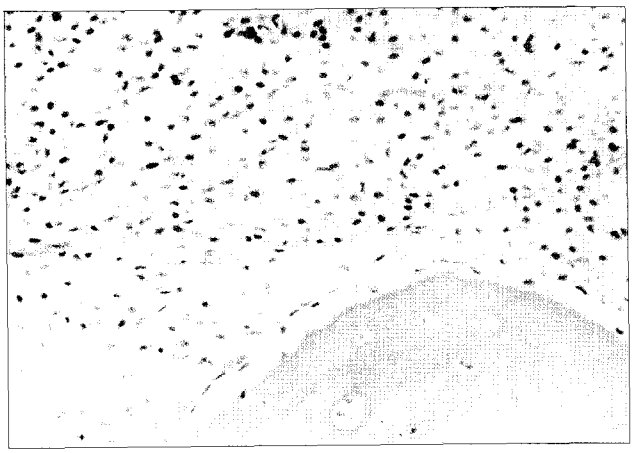

Fig. 4. There is a massive aggregation of histiocytes in the bone marrow (H&E, ×100).

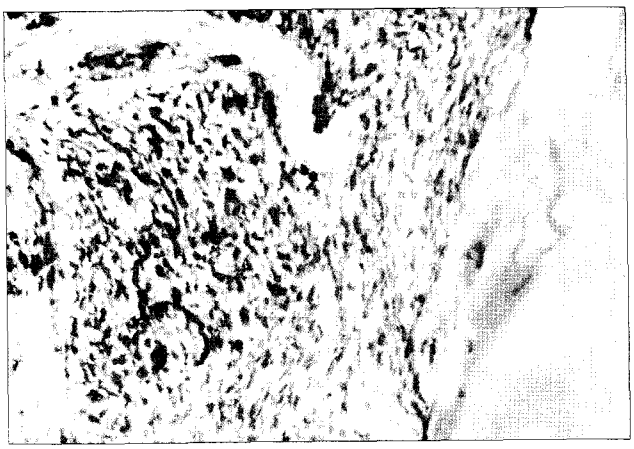

Fig. 5. The histiocytes are stained by an immunohistochemical staining for CD 68 immunohistochemically (CD 68, ×100).

of the histiocytes around the neoplastic plasma cells (Fig. 4). The histiocytes were positive for vimentin and CD68 (Fig. 5).

A post operative systemic survey indicated solitary nature. After surgery, the clinical symptoms improved (lower extrimities Motor Gr IV$^+$, under T7 sensory dermatome 80%). three weeks later he received radiation treatment (total 40Gy). After surgery serum protein electrophoresis revealed monoclonal gammopathy of undetermined significance(MGUS). However, laboratory and chromosomal examination of the bone marrow aspiration from the iliac bone was normal. The final pathological diagnosis was a solitary plasacytoma.

Discussion

Solitary plasmacytoma of bone accounts for only 3% of all plasma cell tumors and consists of one or possibly two lesions. There is a 2-3 to 1 male-to-female predominance, and the average age of the patients is more than 50 years. Between one quarter and one half of all lesions occur in the spine. The thoracic spine is a common location, which is followed by the lumbar spine, cervical spine and sacrum. However, the

initial diagnosis may be difficult to differentiate from an aneurysmal bone cyst, giant-cell tumor, hemangioma, or metastatic spine tumor[5]. The clinical presentation is local pain or the effect of a root or cord compression[1], and coagulopathy due to paraproteinemia can infrequently occur. Radiology reveals a lytic lesion and focal end-plate fractures, possibly with a cystic component. Sclerosis may also be present. The posterior elements of the spine are rare site of origin. However, it can appear everywhere in the three colum[9]. Involvement of the intervertebral disc and adjacent vertebrae is rare. Immunoelectrophoresis of the urine and serum shows a monoclonal band, which resolves after definitive treatment.

Bone scanning and skeletal survey radiography are used to rule out an additional lesion. MRI is a sensitive imaging tool for detecting additional lesion[1,5,8]. Initially, a needle biopsy of the lesion as well as a bone marrow biopsy should be performed to rule out multiple myeloma.

The criteria for a diagnosis of SPB include (a) a solitary bone lesion, (b) bone marrow plasmacytosis less than 10%, (c) biopsy evidence of plasma cell neoplasm and (d) no evidence of other lesions based on the clinical examination or skeletal surney. Our case met all these criteria[2].

Radiotherapy is the first line of treatment for lesions not presenting with rapid neurological deterioration[6]. A local recurrence is rare when more than 45Gy is given. Local control with radiotherapy is excellent, with cure rates up to 96% quoted in the literature and survival up to 11 years, Approximately half of all patients progress to a myeloma 5 years after initial diagnosis[3].

A surgical resection is performed for those lesions causing neural compression and significant neurological deterioration or bony instability. A complete resection and reconstruction with a bone graft and instrumentation are the goals in plasmacytoma so that a cure can be achieved[1]. The conventional anterior route surgery, combined with radiation and chemotherapy is the treatment of choice. However a tumor involving both the anterior and posterior columns can be resected in one-session via the posterior route total *en bloc* spondylectomy for the complete resection of a solitary palsamacytoma. A subtotal tumor resection should be followed by radiotherapy and possibly chemotherapy. A resection of as much of the tumor as possible may lower the need for chemotherapy and reduce the radiation dose needed.

The paraprotein levels should be followed rule out a recurrence or dissemination[10], and the presence of an immunoglobulin M component has been used to predict progression to myeloma[3].

Gaucher disease refers to a cluster of autosomal recessive disorders resulting from mutations at the glucocerebrosidase locus on chromosome 1q21. The affected gene encodes glucocerebrosidase, an enzyme that normally cleaves the glucose residue from ceramide. As a result, glucoscerebroside accumulates principally in the phagocytic cells of the body but in some forms also in the central nervous system. The distended phagocytic cells, known as Gacuher cells, are found in the spleen, liver, bone marrow, lymph node, tonsils, thymus, and Peyer patches. Gaucher cells rarely appear vacuolated but instead have a fibrillary type of cytoplasm likened to crumpled tissue paper. Gaucher cells are often enlarged, sometimes up to 100㎛ in diameter, and have one or more dark, eccentrically placed nuclei. Periodic acid-Schiff(PAS) staining is usually intensely positive. with the electron microscope, the fibrillary cytoplasm can be resolved as elongated, distended lysosomes, containing the stored lipid in stacks of bilayers[4,7]. Pseudo-Gaucher cells are also called Gaucher-like cells, and it found in the bone marrow, spleen and the other site without clinical evidence of inherited Gaucher disease[4,7,10].

Conclusion

A Solitary plasmacytoma of the bone(SPB) in the thoracic spine is a very rare lesions. Accurate preoperative diagnosis may be difficult. However, MRI studies with CT scan may help in the preoperative diagnosis. If a SPB is diagnosed, total surgical removal should be considered to achive a cure and reduce recurrence.

Also, more basic pathological and hematological research works should be contimuously conducted in the future.

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