Solitary plasmacytoma of bone – a rare disorder with an unusual evolution

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Summary: A 40 year old woman presented with a spinal epidural tumour, which on histology was shown to be a plasmacytoma. At that time she had no evidence of multiple myeloma. Ten months later, she developed a second isolated plasmacytoma in the spleen, for which she underwent splenectomy. Two years after her initial presentation she had another recurrence in the liver, followed by a full-blown picture of multiple myeloma. The myeloma was progressive and resistant to all forms of chemotherapy. She finally died of a massive gastrointestinal haemorrhage. The clinical features, natural evolution and management of solitary plasmacytomas are discussed.

Introduction

Localized plasmacytomas in the form of solitary plasmacytoma of bone (SPB) or extramedullary plasmacytoma (EMP) are uncommon, and constitute less than ten per cent of all plasma cell neoplasms.¹ The overall prognosis and survival of the localized plasmacytomas is much better than that of overt myeloma, with complete recovery in some, after adequate local therapy.^{1–3} We report an unusual evolution of this rare form of plasma cell dyscrasia, where a SPB recurred as an EMP, and then developed an aggressive form of disseminated myeloma, resulting in the death of the patient.

Case report

A female aged 40 years presented with a 3 month history of low backache. X-rays showed narrowing of L4-5/S1 disc space. Computed tomographic (CT) scan showed an area of destruction of the right sacrum and a pre-sacral lesion or spinal tumour was suspected. Following a hemilaminectomy at S1–S2 level, a epidural tumour was removed. Initial histology reported this to be a highly anaplastic tumour, probably a secondary carcinoma of unknown primary. She was given radiotherapy to the sacral area. Subsequent review of the histology, however, showed the tumour to be a plasmacytoma. The serum immunoglobulins, protein electrophoresis and bone marrow did not reveal any abnormality. A diagnosis was made of a solitary plasmacytoma of bone.

Ten months later, on follow-up, she was observed to have a solitary, enlarging, lesion in the spleen. Histology, following splenectomy, confirmed an isolated extramedullary plasmacytoma of the spleen. The bone marrow remained normal and she still had no evidence of disseminated myeloma.

Fifteen months after the first recurrence, she developed hepatomegaly. CT scan showed a 12 × 8 cm mass in the liver.

Liver biopsy confirmed the diagnosis of hepatic plasmacytoma. She subsequently developed disseminated myeloma of IgG kappa type. The bone marrow revealed a plasmacytosis of 10%. Serum monoclonal band (M band), IgG type, was quantified at 17 g/l with suppression of other immunoglobulins.

As the patient was young, in reasonably good health, and with normal renal function, she was initially treated with combination chemotherapy (vincristine, adriamycin and methyl prednisolone). She received two cycles, but developed severe bradycardia on both occasions, and was therefore given two cycles of melphalan and prednisolone. The myeloma was resistant to both these regimens, so she was treated with six cycles of vincristine with oral etoposide, given at 3–4-weekly intervals. Following the first three cycles she felt better, and there was a definite regression of the liver mass, as observed on ultrasonographic evaluation. The liver function also improved and the M band was static at 13 g/l. This response, however, was short lived, and she deteriorated over the next few months, finally developing thrombocytopenia, epistaxis and a gastrointestinal haemorrhage from which she died.

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Post-mortem examination revealed widespread metastatic plasmacytoma. Multiple viable tumour masses were observed within the right lobe of the liver. In addition, necrotic yellow tumour was also present within the liver which on histological examination showed large areas of scarring with macrophages containing haemosiderin, consistent with resolved tumour. Some nodules were entirely necrotic with no viable tumour cells present. There was thus evidence of response at some sites, but progression at others.

Discussion

Solitary plasmacytoma of bone is a rare tumour and represents only 3% of all plasma cell malignancies.⁴ A strong male preponderance of 3:1 is reported, in contrast to multiple myeloma, where the sex ratio is close to one.⁵ The mean age of around 51 years for SPB is about 10 years lower than that for indolent or overt myeloma.^{1,6} The usual presentation is with bone pain; however, 25% develop neurological dysfunction in the form of cord compression or nerve root disorder.¹ In SPB, more than half (53.3%) of lesions involve the spine, compared with only 46% in the peripheral bones.⁷

The natural evolution over a 10 year follow-up period illustrates that 85% of cases of SPB either have a local recurrence (12%), give rise to a new solitary lesion (15%) or develop typical multiple myeloma (58%).¹ Dissemination nearly always occurs in the first 3–5 years, and is more commonly seen with primary lesions in the spine (61%) than with lesions in peripheral bones (26%).^{7–9} Our case,

however, had an unusual evolution, passing through the stages of SPB with recurrences as EMP, first in the spleen and then in the liver, finally progressing to myeloma resistant to therapy. The progression of SPB to an EMP in spleen is very rare. It is most unusual for an EMP to produce two sites of extramedullary deposit, in the spleen and liver, on two different occasions in the same patient.

The treatment of choice for SPB is radiotherapy, 4,000 cGy in daily fractions over 3–4 weeks.¹⁰ Following vigorous staging, long-term follow-up of patients with SPB treated with between 3,000 and > 5,500 cGy, showed, on linear trend analysis, no clear relationship between local dose and long-term stability or the onset of multiple myeloma.³ Surgical excision, if possible, may be undertaken, especially with lesions in the spine. Local surgery is usually followed by radiotherapy. Optimal treatment for epidural plasmacytoma with cord compression is laminectomy followed by radiotherapy.⁹ EMPs are radiosensitive, and radiotherapy remains the treatment of choice.⁸ However, as reported here, surgical excision in the form of splenectomy for EMP of the spleen is an appropriate alternative. Chemotherapy is used to treat patients with SPB only when progression to multiple myeloma is evident.

The median survival of SPB is reported to be greater than 10 years.¹ The prognosis of SPB with subsequent dissemination is also considered to be better, with longer survival than for presentation with overt myeloma.² Unfortunately, this case did not behave in the usual manner, rapidly developing an unresponsive tumour.

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