Pediatric Solitary Plasmacytoma

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Solitary plasmacytoma of bone (SPB) is a localized plasma cell neoplasm. SPB accounts for 5-10% of all plasma cell dyscrasias (1-3). The relationship of SPB to multiple myeloma continues to be controversial. Many authors believe that SPB is an early stage of multiple myeloma (1, 3-5), whereas others consider it as a distinct clinical entity (6, 7). The occurrence of SPB in younger age groups and involvement of long bones is very rare. The youngest patient reported in the literature is 16 years old. Here we report a 15-year-old girl with SPB of the humerus with 10 years follow-up. She still remains in remission.

Case report. A 15-year-old girl presented in March 1986 with a history of left shoulder joint pain of 6 months' duration. Pain appeared mainly on lifting the limb above the head. There was no history of any trauma or swelling of the joint. On examination she was moderately built and nourished. There was no pallor or lymphadenopathy and no tenderness or swelling of the joint. Movements of the joint were full. A healed scar was seen over the anterior part of the left shoulder. Systemic examination did not reveal any abnormality. Suspecting a bone tumor open biopsy with curettage and bone grafting was performed in a peripheral hospital. She was later referred to us for further management.

Investigations revealed Hb of 135 g/l. TLC of 7.5×10^9/l, platelets 300×10^9/l, and ESR 40 mm/1st hour. Urine analysis was normal. An x-ray of the shoulder joint showed osteolytic areas with ill-defined borders in the medial part of the head of the humerus (Figure). The biopsy showed mainly plasma cells in an amorphous background of which some were binucleate but the majority were mature. A few immature plasma cells were noted. In view of the plasma cell neoplasm she was investigated further for evidence of any systemic disease. However, skeletal survey, serum electrophoresis, immunoglobulin quantitation, bone marrow aspiration and biopsy were all normal. Bone scan showed a solitary lesion in the left humerus. In view of the isolated plasma cell tumor the diagnosis of solitary plasmacytoma of the bone was considered. She was treated with local radiotherapy—42 Gy in 14 fractions. The treatment was well tolerated. She has been followed up for the last 10 years and is still in remission.

Discussion. Plasma cell neoplasma may be either localized (solitary plasmacytoma) or generalized (multiple myeloma). Solitary plasmacytoma may arise either in bone (solitary plasmacytoma of bone—SPB) or in extramedullary tissues. SPB accounts for 5-10% of patients with plasma cell neoplasm (1-3).

The median age at presentation with SPB is 50 years, 7-10 years less than that of patients with multiple myeloma (1, 4, 6, 8). The youngest patient reported was 16 years old (8). To the best of our knowledge only 2 cases have been reported below the age of 20 (3, 8) and 70-90% of the patients are males (3, 6). SPB may involve any bone but is most commonly seen in the axial skeleton, mostly in the vertebrae (40-60%) (8), where the thoracic vertebrae is involved most frequently. Long bones are rarely involved, femur and humerus accounting for 9% of the cases each in the series by Holland et al. (5) and Meis et al. (9) and 5.5% in the series by Bataille & Sany (6). A slightly higher incidence (20%) was noted by Knowling et al. (1). Criteria for the diagnosis of SPB include a) solitary bone lesion, b) bone marrow plasmacytosis lessthan 10% c) biopsy evidence of a plasma cell neoplasm and d) absence of evidence of other lesions based on clinical or skeletal survey (6). Myeloma protein is present in the serum and urine in a significant number of patients with a reported incidence varying from 24 to 67% in different series. However, unlike in multiple

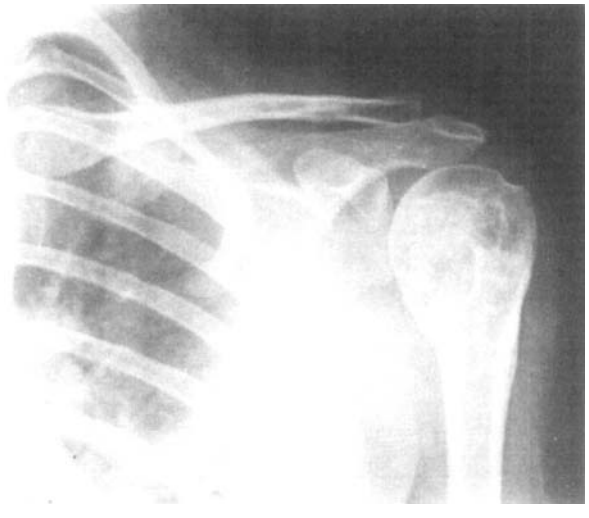

Figure. Radiograph of the humerus showing osteolytic area with ill-defined borders.

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myeloma uninvolved immunoglobulins are normal in more than 90% of patients.

The mainstay of treatment is local radiotherapy. The amount of radiotherapy given varies from 35 to 50 Gy, but there is no clear dose–response relationship. Myeloma protein disappears with local radiotherapy in 25–50% of patients (1, 4, 6, 8).

The opinion regarding the role of adjuvant chemotherapy in SPB is divided. A few studies have shown that adjuvant chemotherapy could delay the progression to multiple myeloma with prolongation of overall survival, whereas others have failed to demonstrate any benefit (6, 10). According to Bataille & Sany (6) a better prognosis was seen in patients with peripheral than with spinal solitary plasmacytoma. Frassica et al. (8) and Shih et al. (10), however, could not find any such association.

The overall survival is much better than that of multiple myeloma. Progression to overt myeloma occurs in approximately 50% of patients and new bone lesions or local recurrence develop in about 10% of the cases (8). 10 years of disease-free survival was about 25% (4, 8) previously, but with the advent of newer investigations to exclude systemic disease the 10-year survival has increased to 42% (2).

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