CASE REPORT

Ewing's Sarcoma of the Talus in a Four-Year-Old Child

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Summary

Ewing's sarcoma is a rare malignant tumor of the foot in children. We report a case of Ewing's sarcoma of the talus in a four-year-old Chinese girl to highlight the initial difficulty in diagnosis and the clinical course of the disease. She was initially diagnosed as osteomyelitis of the talus and died eight months after presentation with pleural and spinal metastases. To the best of our knowledge, Ewing's sarcoma of the talus in a young child has never been reported in Malaysia.

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A four-year-old Chinese girl was admitted to our hospital in June 1999, with a history of recurrent right ankle pain, swelling and low-grade fever of five months duration. In March 1999, she had a skin infection over her right ankle and was treated with antibiotics by a general practitioner. She had a normal perinatal history and had completed her immunization. There was a strong family history of malignancy. Her grandfather, grandmother and uncle had died from nasopharyngeal carcinoma, breast carcinoma and hepatoma respectively.

Clinically, she was a febrile and of average built. The gait was antalgic. There was a firm, mildly tender, diffuse swelling on the lateral aspect of the right ankle. The range of ankle movement was normal. The right inguinal lymph node was palpable and measured 1cm in diameter. The liver and spleen were not enlarged. Both lungs were normal. The total white count was normal and the erythrocyte sedimentation rate was 46 mm/hour.

The plain x-ray was reported as osteomyelitis of the right talus (Fig. 1). She was treated empirically with antibiotics and analgesics and subsequently discharged. Her ankle swelling and pain were reduced at follow-up one week later.

One month later, she was readmitted in another hospital with a cough and progressive swelling of the right ankle. The CT scan of the thorax showed a metastatic lesion and an effusion in the left pleural cavity. A biopsy of the right ankle showed small synovial villous nodules consisting of local proliferation of endothelium. There were no malignant cells seen.

She was transferred to our hospital when the left pleural effusion worsened. A chest tube was inserted and her condition improved in the intensive care unit. A repeat biopsy of the talus and left pleural were done after a few days when her general condition improved. Intra-operatively, the right ankle showed a friable whitish tissue infiltrating the synovium and talus.
Fig. 1: X-ray of the right ankle showing sclerosis of the talus.

Histopathological examination confirmed an Ewing's sarcoma. Microscopically, the tumour consists of small round cells. The cells react with MIC2 antibodies. The leucocyte common antigen was negative and the actin, desmin and myoglobin markers were absent (Fig 2 a and b). She was referred to the oncologist for radiotherapy and chemotherapy. The pleural effusion resolved dramatically within 5 days of radiotherapy. Combination chemotherapy (consisting of ifosfamide, vincristine and adriamycin) was commenced.

During chemotherapy four months later, she developed an acute onset of paraplegia. A bone scan did not show any increased uptake in the spine but the MRI of the spine showed multiple metastases to the thoracic and lumbar spine with extradural compression of the spinal cord at the level of the second and third lumbar spine. She had radiotherapy to the spine but there was no neurological recovery.

Fig. 2 A: Photomicrograph of the ulnar lesion showing small round cell infiltrate (hematoxylin and eosin x 200).

Fig. 2 B: Photomicrograph of the small round cells reacting positively with MIC 2 antibodies (immunoperoxidase x 200).
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In February 2000 she received another course of chemotherapy. Adriamycin infusion was given over one hour, followed by ifosfamide infusion over 3 hours. After two hours of intravenous ifosfamide, she died suddenly, most probably due to a cardiac arrest.

Discussion

Ewing's sarcoma is a malignant, non-osteogenic primary tumor of bone. James Ewing in 1921 first reported a 14-year-old girl with a lytic lesion of the ulna that responded dramatically to radiotherapy (cited in Vlasak and Sim, 1996). Ewing's sarcoma in young children is rare, highly malignant, and when at an unusual site it is often difficult to diagnose. Because the prognosis of metastatic Ewing's sarcoma remains poor, early detection is the best hope for long-term survival. Durbin et al. noted that the delays in the diagnosis of Ewing's sarcoma in young children were due to several reasons.

Firstly, since most bony lesions in this age group are benign, malignancy would be low in the list of differential diagnosis. Secondly, only 8% of all childhood malignancies occur in the bones and Ewing's sarcoma accounts for only 25% of these bone malignacies, the remainder being mainly osteosarcoma or chondrosarcoma. Thirdly, only 3% of Ewing's sarcoma occurs below the age of three as the majority of patients are in their second decade of life. Fourthly, the diagnosis may be overlooked when the lesion occurs in an uncommon site as the talus, instead of the long bones, pelvis, or ribs.

Adkins et al. reported a marked delay in diagnosis when it occurs in the hind-foot (mean, 22 months) rather than in the forefoot (mean, 7 months). There is little information specifically addressing Ewing's sarcoma of the talus. Weissman et al. in 1966 (cited in Adkins et al., 1997) reported an 18-year-old girl with an osteosclerotic tumor in the talus that initially was treated for avascular necrosis. Pandey in 1970 (cited in Adkins et al., 1997) similarly reported a patient with an osteosclerotic talar lesion who was treated for avascular necrosis.

In addition, Ewing's sarcoma can simulate an infection hence delaying the diagnosis. The symptoms of pain, which may be intermittent as in this patient is often misleading and especially so when it is associated with swelling and fever and could be mistaken for osteomyelitis.

A definite diagnosis of Ewing's sarcoma was made based on special staining and immunohistochemical studies. This was not done for the first biopsy. Our patient had metastases to the lungs and spine. Pritchard et al. (cited in Durbin et al., 1998) found that 25% of Ewing's sarcomas have metastasized at the time of diagnosis. For non-metastatic Ewing's sarcoma the 5-year, disease free survival is 70%, whilst for metastatic disease, it is only 33%. Adriamycin may have caused cardiac toxicity in our patient.

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References
