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Solitary cerebellar metastasis from Ewing's sarcoma: case report and review of the literature

Meng-Fai Kuo, Swei-Ming Lin, Yong-Kwang Tu

Department of Surgery, National Taiwan University Hospital, 7, Chung-Shan S. Road, Taipei, Taiwan 10016, Republic of China

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Abstract. A rare case of Ewing's sarcoma metastatic to the cerebellum is presented. Neurosurgical intervention was required which played a significant role in the treatment of this patient. The incidence and treatment of central nervous system involvement from Ewing's sarcoma is reviewed and discussed.

Key words: Ewing's sarcoma – Cerebellar metastasis

Introduction

Ewing's sarcoma is an uncommon malignant neoplasm of bone which accounts for approximately 10% of all malignant primary bone tumors. This tumor mainly affects children and young adults [2, 9] and originates most often in the long bones of the lower extremities and pelvic girdle [1, 2]. In 20% of cases, metastatic lesions are present at the time of initial diagnosis [1]. Metastasis occurs in up to 85% of patients within 2 years of diagnosis [4], generally involving the lungs and the skeletal system [1]. Central nervous system (CNS) metastasis has been reported in 32–56% of cases and often reflects meningeal invasion by direct bony extension [6, 8, 9]. Intracerebral metastasis, however is distinctly uncommon, occuring in only 1.8% of cases [16]. Improved long-term survival secondary to refined adjuvant chemotherapy and satisfactory control of local recurrence by limited field irradiation or surgical treatment are thought to contribute to the increased incidence of CNS lesions [1, 5, 12]. The blood-brain barrier is thought to be impermeable to adjuvant chemotherapeutic agents, resulting in failure to eliminate microscopic CNS metastases [6].

We report a case of Ewing's sarcoma with a solitary cerebellar metastasis that was successfully removed. A metastatic tumor in this site has not been previously reported. The possible role of neurosurgery in the treatment of this condition is discussed.

Correspondence to: M.-F. Kuo

Case report

A 12-year-old girl developed pain in the left thigh 6 months prior to her first admission to National Taiwan University Hospital. Three months later she began having local tenderness, swelling, and a limping gait. Roentgenography showed mottled bone destruction in the left femoral shaft. The tumor elevated the periosteum, producing an "onion skin"-like subperiosteal reaction (Fig. 1). She then underwent an open biopsy for the femoral lesion. The pathology analysis showed a Ewing's sarcoma. She was treated with local irradiation (5000 rads) and chemotherapy with vincristine, adriamycin, actinomycin D, and cyclophosphamide for the next 15 months. Pathological fracture of left femur occurred at the end of chemotherapy. An open biopsy revealed fibrosis and irradiation effect, so the femoral shaft was resected and intramedullary vascularized fibular grafting was carried out.

The patient did well for 14 months after chemotherapy until she was readmitted to the hospital because of persistent vertigo and vomiting. Neurological examination revealed dysmetria, dysdiado-

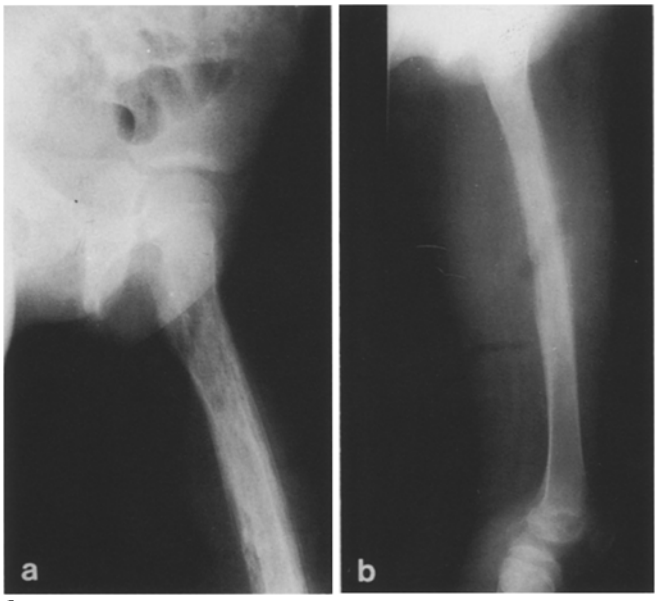

Fig. 1a, b. Left femoral roentgenography shows destruction of the bone and "onion skin"-like subperiosteal reaction. **a** Anteroposterior and **b** lateral views

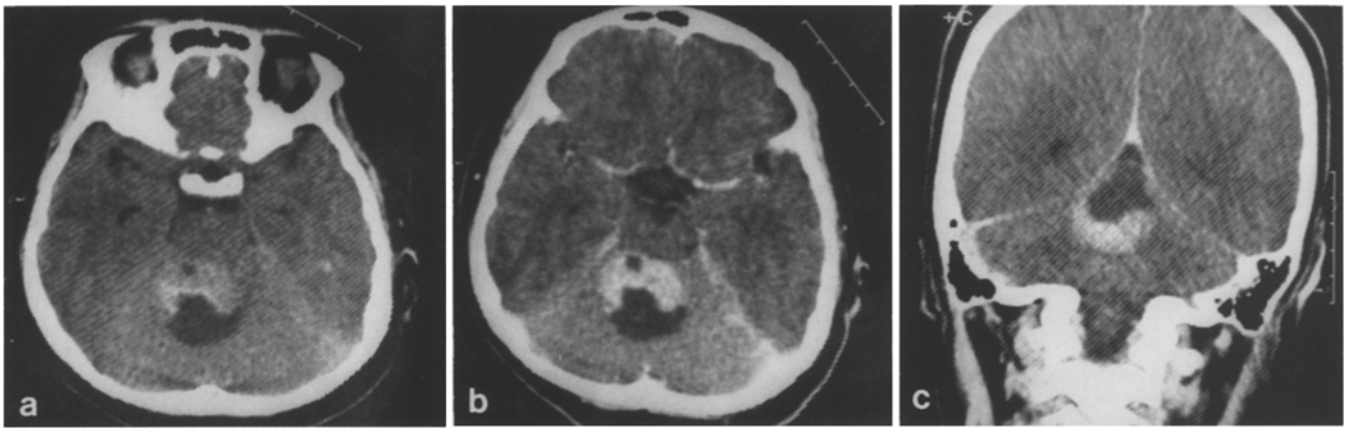

Fig. 2a–c. Brain computed tomographic scan shows a slightly hyperdense mass comprising cystic and solid components in the vermis. After intravenous administration of contrast medium, the solid part enhanced well. **a** Precontrast axial scan, **b** postcontrast axial scan, **c** postcontrast coronal scan

chokinesia, and a positive Romberg's sign on the right. A brain computed tomographic scan revealed a well-defined contrast-enhanced lesion comprising cystic and solid components in the midline portion of the posterior fossa. The IV ventricle was compressed and mild hydrocephalus was noted (Fig. 2). The patient underwent suboccipital craniectomy for gross total removal of the tumor. The tumor was fragile and cystic. The cyst contained clear, colorless fluid. There was a discrete margin between tumor and normal cerebellum. The overlying tentorium was not involved. The histology was consistent with the diagnosis of Ewing's sarcoma and similar to the initial biopsy obtained from the left femur.

The postoperative course has been uneventful to date (16 months). Computed tomography, magnetic resonance imaging, bone scanning, and chest X-ray follow-up showed no evidence of tumor. The cerebrospinal fluid study was unremarkable. A bone marrow biopsy was negative. The patient is still under regular follow-up.

Discussion

Ewing's sarcoma, initially described by James Ewing in 1921, is a highly malignant tumor of bone. In his comprehensive review in 1941, CNS involvement was not mentioned. Prior to the advent of adjuvant chemotherapy, the treatment for primary Ewing's sarcoma mainly depended on surgical excision and local irradiation, which gave very low 5-year survival rates of less than 10% [2, 3]. Using aggressive therapy including radiotherapy and chemotherapy, Pomeroy and Johnson reported a 35% 5-year survival [11]. Since the use of a five-agent chemotherapeutic protocol consisting of serial intravenous administration of vincristine, actinomycin D, adriamycin, cyclophosphamide, and decarbazine [15, 16], 5-year survival rates of approximately 50% have been reported [15]. The success of these therapeutic modalities has been suggested to be responsible for the increasing incidence of CNS metastases from Ewing's sarcoma [8].

The most common form of CNS involvement by Ewing's sarcoma is neural compression and secondary involvement of the meninges by direct extension of the adjacent affected spine or skull. True metastasis to the nervous tissue is extremely rare. The incidence of metastasis to the CNS in cases of Ewing's sarcoma ranged from 2.2% to 19% [7, 9, 16]. In 1970, Kulick and Mones [7] presented a series of 100 patients with Ewing's sarcoma. In this group, 32 patients were found to have neurologic involvement. However, only one patient had proven intraparenchymal CNS Ewing's sarcoma. A study by Marsa and Johnson in 1971 [8] presented 20 patients with localized Ewing's sarcoma. Two patients developed intraparenchymal and meningeal involvement, in one case proved by autopsy.

In 1974, Mehta and Hendrickson found neurologic symptoms in 15 of 27 patients. Four of them had evidence of intraparenchymal Ewing's sarcoma without bony or dural involvement on brain scan. In only five patients was confirmation obtained by autopsy, but all demonstrated extensive dural metastasis [9]. Kies and Kennedy [6] confirmed CNS metastasis in 13 patients (12%) by autopsy. Only three patients (2.2%) had discrete intraparenchymal brain metastasis. Intraparenchymal or meningeal metastasis without adjacent bony or dural involvement was found on only 10 patients out of a group of 445 (2.2%) entered into protocols of the National Cancer Institute and Intergroup Ewing's Sarcoma Study [16]. Eight (1.8%) of these patients had cortical lesions, and the other two (0.4%) had meningeal involvement. In Vannucci and Baten's series [18], 231 brains in pediatric patients with malignant tumors were examined. Excluding 14 cases of primary brain tumor, the overall incidenc of metastatic brain tumor was 6%. None of the 15 patients with Ewing's sarcoma had cerebral metastasis.

Surgical decompression of the spinal cord and the brain from bony or dural extension of this disease has been described repeatedly [6, 19], but only three cases of intracerebral Ewing's sarcoma treated by surgical removal have been reported [10, 13, 14]. All of the CNS intraparenchymal metastases from Ewing's sarcoma reported in the literature were in the supratentorial area [4, 6–8, 12, 17], and only 16 histologically proven cases of intracerebral lesions have been documented [1, 6, 8, 10, 13, 14, 16]. A metastatic Ewing's sarcoma in the cerebellum that was surgically removed has not been reported before.

With longer survival in patients with Ewing's sarcoma, intracerebral metastases will occur with increasing frequency [6, 14, 16, 17]. The influence of neurosurgical intervention on the outcome has yet to be determined, because on the small number of operative cases reported. However, surgical removal of these lesions must be considered as an integral part of treatment.

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