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ATYPICAL CASE OF METASTATIC EWING SARCOMA WITH DISEASE STABILISATION AND POST-CHEMOTHERAPEUTIC GANGLIONEUROBLASTOMA-LIKE DIFFERENTIATION

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INTRODUCTION

Ewing sarcoma is a highly aggressive malignancy, typically affecting people aged 10-20 years, with a 5-year survival rate for metastatic disease of less than 30%. Uncommonly, following chemotherapy, the disease can stabilise and even differentiate into a ganglioneuroblastoma-like tumour.

AIM

To report a case of Ewing sarcoma with unusual disease development.

METHODS

Our patient is a 16-year-old male, diagnosed with EWSR1-ERG gene fusion Ewing sarcoma, involving a large primary tumour in the right iliac bone, along with metastases in the liver, lungs, 10th thoracic vertebra and mediastinal lymph nodes. As neoadjuvant treatment, the patient received 1 VIDE (vincristine, ifosfamide, doxorubicin, etoposide) cycle followed by 8 alternating cycles of VDC/IE (vincristine, doxorubicin, cyclophosphamide / ifosfamide, etoposide) at 14-day intervals, followed by 12 irinotecan/temozolomide (IT) cycles. During chemotherapy, the patient underwent radiotherapy, fractionated into 30 sessions of 1.8 Gy up to a total dose of 54 Gy. MRI and CT scans were used for initial and follow-up radiological evaluation and PET scan for metabolic activity evaluation.

RESULTS

Neoadjuvant treatment had a poor effect on both the primary tumour and the metastases (minimal regression on radiological evaluation, primary tumour remained unresectable, all lesions hypermetabolic on PET scan). On subsequent rebiopsy, the primary tumour had a very low proliferative index (Ki67 < 5%). After 12 IT cycles, albeit seemingly stable on radiological evaluation, the lesions displayed decreased metabolic activity on PET scan. Moreover, the primary tumour revealed central necrosis on MRI (Figure 1), while the biopsy of the liver metastasis showed a ganglioneuroblastoma-like differentiation.

CONCLUSIONS

Treatment of metastatic Ewing sarcoma remains a major challenge. Nevertheless, even with poor response to chemotherapy, metastatic Ewing sarcoma can transition to a progression-free and stable state. Further research into the treatment of Ewing sarcoma is crucial, especially given the neural maturation findings, whose prognostic significance remains unknown.

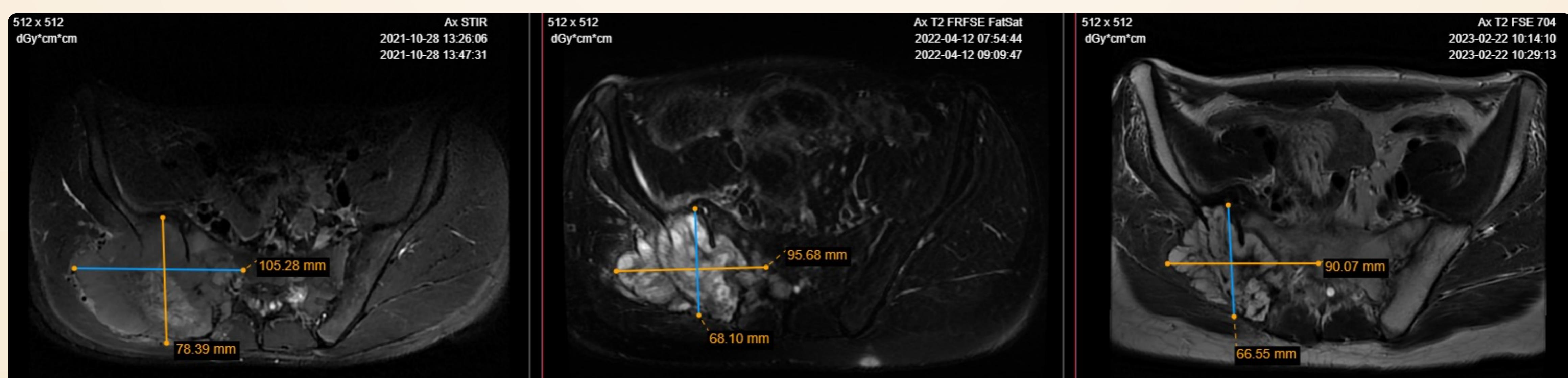


Figure 1. From left to right - initial MRI findings of the primary Ewing sarcoma tumour, control MRI after neoadjuvant treatment, latest control MRI after radiotherapy and 12 irinotecan/temozolomide cycles.