

# CASE 004

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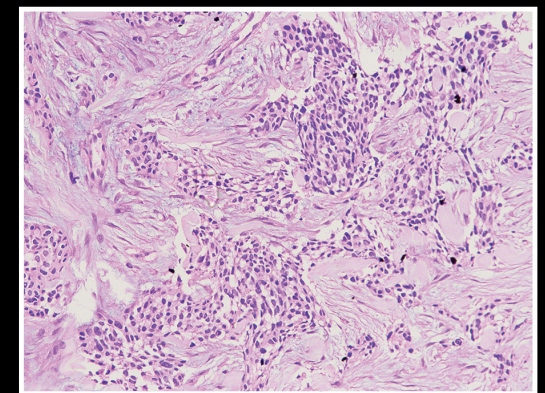
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## INTRODUCTION

Ewing's sarcoma (ES) is the second most common bone tumor after osteosarcoma in children and adolescents. ES are aggressive tumors with a tendency towards recurrence following resection and pronounced proclivity toward early hematogenous metastases to lungs and bone. Although it is the second most common bone cancer seen in children, it is relatively uncommon and accounts for only 1% of all childhood cancer cases. It belongs to a group of tumors often called Ewing's family of tumors. Two other distinct entities namely the primitive neuroectodermal tumors (PNETs) and the Askin's tumour of the chest wall are also included in this family of tumors. No hereditary or congenital syndromes, environmental, or known risk factors have been associated with the occurrence of ES. In 90% of cases, Ewing's sarcoma family tumor (ESFT) cells harbor the translocation  $t(11;22)(q24;q12)$ , and in the remaining 10% the variant translocation is  $t(21;12)(22;12)$ <sup>1,2</sup>. Although peak incidence occurs between the ages of 10 and 20 years, patients of younger or older ages account for almost 30% of the cases<sup>3</sup>. Poor prognostic factors include tumor  $\geq 8$  cm, pelvic primary, presence of metastasis, and age  $>15$  at the time of diagnosis<sup>4</sup>. Older patients with sarcoma also have a higher risk of thromboembolism.

## CASE REPORT

A 23 year old male presented with abdominal pain, loss of appetite, and on examination revealed a pelvic mass. There was no family history of malignancy or any other significant past history of any illness. A trucut biopsy of the large pelvic mass pressing on the bladder and colon, reaching the prostate, but not infiltrating and well delineated, was done. Grossly there were four linear bits of tissue measuring 0.5 cm to 1 cm in length. Microscopy showed islands of small round cells with hyperchromatic nucleus and spotty necrosis. Few cells were spindled and showed clear cytoplasm. Mitosis was present. After running a panel of immunomarkers, a diagnosis of Desmoplastic round cell tumor was given. This biopsy was not available for review at CORE Diagnostics. The patient received



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