

Ollier's disease associated with ovarian juvenile granulosa cell tumor and triple X syndrome: A letter to the editor

Dear Editor,

Ollier's disease, known as multiple enchondromatosis, is a rare nonhereditary benign bony lesion, which can be found in early childhood. Enchondromas frequently are developed in long tubular bones and flat bones. The lesions may affect multiple bones and are usually asymmetrically distributed, predominantly affecting one side of the body.^[1] Radiologic features demonstrate multiple radiolucent, homogeneous density lesions with an oval or elongated shape and well-defined slightly thickened bony margins.^[2,3] The association of Ollier's disease and malignant transformation to chondrosarcoma has been described in 30%–50% of cases. Pancreatitis, anaplastic astrocytoma, and liver cancer have been reported, with less commonly.^[4,5] Malignant ovarian neoplasms and their association with Ollier's disease have rarely been described in the literature.

A 22-month-old female infant was referred to the orthopedic department of our hospital with left leg deformity and discrepancy in lower limb length. Radiologically, multiple lower limb and right upper limb enchondromas were found, and a diagnosis of Ollier's disease was established [Figure 1]. She was preterm, delivered at 32 weeks' gestation. A karyotype study was performed during the fetal period and triple X syndrome could be detected. At the age of 18 months, sudden progressive ascites were developed. Sonography revealed a large solid cystic mass with multilocular appearance and heterogeneous

echogenicity, as well as internal vascularity. Furthermore, abdominopelvic computed tomography scan imaging revealed an enhanceable heterogeneous mass lesion in the right and central aspect of the abdominal cavity. Laparotomy and histopathological evaluations illustrated irregular-shaped cells in myxoid background, with basophilic secretions and high mitotic count in cell nuclei. Tumor cells expressed cytokeratin, calretinin, ki-67, inhibin, and WT1; a diagnosis of ovarian juvenile granulosa cell tumor (JGCT) was established.

The association between two rare entities, Ollier's disease and JGCT, was reported. Ollier's disease affects unilateral limbs; however, we noted that left lower and right upper limbs could be affected by Ollier's disease. Another interesting aspect of this case report was triple X syndrome (Karyotype 47 XXX), and its association with Ollier's disease and JGCT was established for the first time. The fact that Ollier's disease can increase the risk of various neoplasms should be considered in managing these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.



Figure 1: (a) Left leg deformity and lower limb length discrepancy. Plain radiography of limbs: (b) AP view of pelvis: Bilateral lytic expansile lesions with sclerotic rim in both femoral proximal metaphysis with mostly involvement of intertrochanteric region and greater trochanter. (c) AP view of both knees: Bilateral lytic lesions in both proximal tibial metaphysis and left femoral metadiaphysis causing deformity and shortening of Lt Femur. (d and e) AP and lateral left leg: Lytic lesion in distal metadiaphysis of Lt tibia. (f) AP view of right shoulder: Lytic expansile lesion in right acromial process with sclerotic rim without obvious destruction or periosteal reaction. (g) AP view of the right wrist: Lytic destructive lesion in distal ulnar metadiaphysis with partial sclerotic rim and shortening of the bone; multiple lytic expansile lesions in metacarpal and phalangeal bones with bone deformities and shortening of these long bones

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