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Images in hematology-oncology

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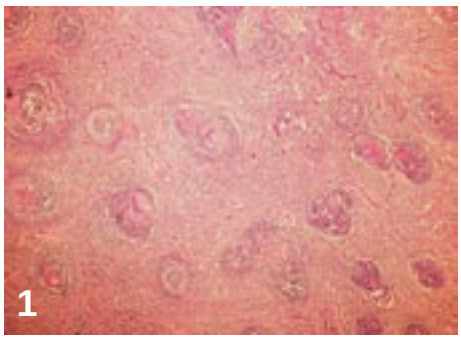
A 14 year-old girl with a cervical mass

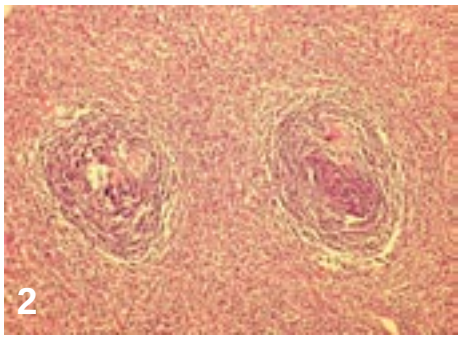
CLINICAL HISTORY

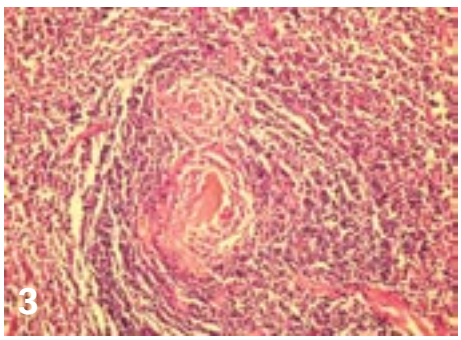
A 14 year-old girl was presented with the complaint of cervical mass. The physical examination was found normal except for a nontender, palpable mass of 4,5x3 cm in diameters on the left cervical area. The blood analysis revealed hemoglobin 14,2 g/dl, hematocrit 37,7 %, leukocyte count 7900/mm^3, platelet count 292.000/mm^3 and blood smear was normal. Other laboratory results were unremarkable. Cervical ultrasonography demonstrated a 5x3,5x3 cm solid hypervascular mass in the left cervical region. Computed tomography (CT) examination of the chest and abdomen were normal. The patient underwent radical tumor resection.

In the gross examination, a solid and homogeneous mass of 4,5x3x2 cm in diameters with a yellow brown colored cut surface was seen.

In the microscopic examination, normal lymph node structure was altered with relatively hypocellular stroma rich in arborizing small caliber vessels. Many small, irregular follicles displaying burnt-out centers were present. A few irregular hyalinized fibrous areas were scattered among the follicles (Figure 1). More than one abortive follicle centers surrounded by one mantle zone lollypop pattern created by hyalinized small vessels entering a follicle perpendicularly, onion skin like arrangement of mantle zone lymphocytes, hyalinized thick vessel walls in the follicle centers were noted (Figures 2 and 3, respectively). No sign for malignancy or specific inflammation was seen.







What is your diagnosis?

PATHOLOGIC DIAGNOSIS

Castleman's disease: Hyaline vascular type

DISCUSSION

Castleman's disease (CD), mainly reported in adults, is a lymphoproliferative disorder of unknown etiology (1). Infrequent case reports are present in children (2). The pathogenesis of CD has not been established yet. CD has been commonly thought to represent a defect in immunoregulation, resulting in an excessive proliferation of B lymphocytes and plasma cells in lymphoid organs (2). Deregulated overproduction of IL-6 has been found to play pathological roles in chronic inflammatory diseases such as rheumatoid arthritis, CD, Crohn's disease and juvenile idiopathic arthritis (3).

Three histologic forms of the disease have been defined: The hyaline-vascular type, characterized by small hyalinized follicle centers and by prominent interfollicular vascular proliferation is the most common. It affects approximately 90% of the patients and usually involves the mediastinum. The plasma cell type, characterized by an abundance of plasma cells, usually involves extrathoracic sites and is less frequent. IL-6 has been produced by the affected lymph nodes and that serum IL-6 positively has been correlated with clinical abnormalities in patients with plasma cell type CD (1,3,4). These patients have systemic inflammatory manifestations such as fever, fatigue, anemia and abnormal laboratory findings such as hyper gamma-globulinemia, hypoalbuminemia, hypocholesterolemia and thrombocytosis. (3,4). In addition to these two types, an uncommon mixed type also exists (2,4).

CD in children is different from the disease in adults mainly because of the rare occurrence of the multicentric forms. The rare occurrence of multicentric forms in children supports the hypothesis that pediatric CD might represent an earlier form where environmental events could play a major role in the development of the disease (1). Surgical excision is curative for the localized variants of CD, either hyaline-vascular or plasma cell type. Most of the patients with multicentric CD are plasma cell type which is a more aggressive clinical entity.

Patients with multicentric CD do not benefit from surgical treatment and for that reason they should be treated by systemic therapy such as steroids and combination chemotherapy (4,5).

Beck et al. treated one patient with murine anti-human IL-6 neutralizing antibodies and they found out some therapeutic benefit (6). It can be suggested that anti IL-6 antibodies may be helpful in the management of this disorder.

References

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