Liver: Nested stromal epithelial tumor

Y Albert Yeh

North Shore University Hospital, Long Island Jewish Medical Center, Hofstra University School of Medicine, New York, USA (YAY)

Identity

Alias
Ossifying malignant mixed epithelial and stromal tumor; Ossifying stromal-epithelial tumor; Desmoplastic nested spindle cell tumor; Calcifying nested stromal-epithelial tumors of the liver

Note
Nested stromal epithelial tumor (NSET) of the liver is an extremely rare non-hepatocytic tumor of the liver and is characterized by nests of spindle and epithelioid cells with occasional calcification and ossification.

Synonyms include ossifying malignant mixed epithelial and stromal tumor, ossifying stromal-epithelial tumor, desmoplastic nested spindle cell tumor, and calcifying nested stromal-epithelial tumors of the liver.

Clinics and pathology

Epidemiology
Twenty four cases of NSET of the liver have been reported. NSET occurs in patients with age range from 2 to 33 years old. The tumor affects mainly children, and females are more frequently affected than males.

Gross and histopathological characteristics of NSET. Grossly, the tumor has a yellow-tan and bulging lobulated appearance. A satellite tumor nodule is present in the periphery (A). Microscopically, nested tumor cells have oval nuclei, stippled chromatin, and inconspicuous nucleoli. The interface between epithelioid and spindle cells is shown (arrows). Courtesy of Sergey V Brodsky.
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**Clinics**
Patients are presented with palpable abdominal tumors. Cushing syndrome has been reported in a few patients.

**Pathology**
Macroscopically, NSETs predominantly occur in the right lobe of the liver and are unencapsulated, well circumscribed tumors range in size from 2.8 to 30 cm in greatest dimension. Satellite tumor nodules also have been reported. Tumors have a yellow-tan and lobulated appearance.

Microscopically, NSETs are characterized by organoid arrangement of cellular nests composed of spindle and epithelioid cells embedded in a desmoplastic or fibrocytic/myofibroblastic stroma, within which proliferation of bile ductules is noted. Areas of myxoid and cystic degeneration or necrosis are sometimes encountered within or adjacent to the cellular nests. Focal psammomatous calcification or osteoid formation is present in some tumors. Lymphovascular invasion is occasionally seen.

The spindle cells within the cellular nests are arranged in short fascicles with a somewhat whorled pattern.

The nested cells are characterized by plump nuclei, stippled chromatin, and inconspicuous nucleoli. Scattered mitotic figures with abnormal forms are identified.

Immunohistochemically, the tumor cells are stained positive for cytokeratin AE1/AE3, keratin CK19 (focal), EMA, CD117 (c-kit), CD56, CD99, ACTH, chromogranin, synaptophysin, neuron-specific enolase, and S100 (focally weak in epithelioid cells). Vimentin stain is positive in the nested spindled cell and stroma. Muscle specific actin and smooth muscle actin immunostains highlight stromal myofibroblastic cells. Alpha-fetoprotein and p53 are negative.

**Cytogenetics**
Chromosomal analysis of one tumor reveals an abnormal karyotype of 60-63,XXX,-1,-4,-5,-others,+2mar.

**Prognosis**
Most patients are doing well with no tumor recurrence in 6 months to 14 years. Tumor recurrence has been observed in two of twenty four patients. One case with aggressive clinical behavior and extrahepatic lymph node metastasis has been reported.

**Immunohistochemical stains of NSET.** Tumor cells are stained positive for immunostains including ACTH (A) (Courtesy of Milton J Finegold), b-catenin (B), cyclin D1 (C), Ki-67 (30% of nuclear staining) (D), p21ras (E), topoisomerase II (F).
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References


This article should be referenced as such: