

## **CASE: HOSPITAL DE S. JOÃO, PORTO, PORTUGAL**

**Presentation: SPAP meeting of May 16, 2012, by Dr. Roberto Silva**

- I. **CLINICAL DATA:** Eighteen year-old female, asymptomatic until 4 months before admission, complaining of abdominal pain. Imaging evaluation disclosed a solid and cystic tumour involving the stomach. The CT scan showed a tumor in the right abdominal quadrant containing cystic areas, septa and multiple foci of calcification, in close contact with the gastric wall. A partial gastrectomy was performed and the surgical specimen displayed a nodular, solid and cystic tumour with 10.5cm in largest dimension.
  
- II. **HISTOPATHOLOGICAL DATA:** On microscopical examination, the tumor was centered in the muscular layer of the gastric wall. It showed a distinct biphasic pattern, composed of epithelial and, predominantly, mesenchymal elements. The epithelial component was arranged in nests, cords and tubules, whereas the mesenchymal component comprised short bundles of spindle cells, foci of multinucleated cells and variable myxoid/collagenous stroma. Tumour cell atypia was mild and the mitotic index was low. There was no evidence of lymphovascular/perineural invasion. Lymph nodes were reactive, without evidence of metastases. Immunohistochemically, epithelial cells expressed AE1/AE3, CAM5.2, CD56 and, focally, CD10; mesenchymal cells expressed vimentin, CD10 and, focally, CD56.
  
- III. **DIAGNOSIS: Gastroblastoma**
  
- IV. **BRIEF HANDOUT:**
  1. **General features. Gastroblastoma:**
    - Very rare epithelial-mesenchymal gastric tumour.
    - Mainly affects young adults and children.
    - Low-grade features with relatively scarce mitotic activity, discrete overall atypia and lack of overt pleomorphism.
    - Low grade of malignancy.
  
  2. **Classification - Biphasic epithelial-mesenchymal tumors of the stomach:**
    - Gastroblastoma**
    - Biphasic synovial sarcoma
    - Teratoma
    - Carcinosarcoma

### 3. Gastroblastoma - Clinical features:

- Young adults and children: range 9–30 years
- Size ranged from 5 to 15 cm, with variable involvement of the gastric wall (either transmural or limited to muscularis propria or serosa)
- Multinodular, partly cystic, with pink-tan to hemorrhagic, yellow-gray cut surface
- Five cases reported (one case with histologically proven nodal metastasis and clinical distant metastases)

### 4. Genesis:

- Unknown
- It has been compared with other blastomas, such as pleuropulmonary blastoma, hepatoblastoma, and nephroblastoma

### 5. Microscopy:

- Biphasic pattern with variable proportion of epithelial and mesenchymal components
- Infiltrative growth pattern
- Epithelial component ranging from primitive-appearing structures intimately associated with the stroma to well-developed luminal structures, with eosinophilic secretions, well demarcated from the stroma.
- Mesenchymal component comprised short bundles of spindle cells, foci of multinucleated cells and variable myxoid/collagenous stroma
- Mild cell atypia and variable mitotic index

### 6. Immunohistochemistry:

	Miettinen <i>et al</i> (2009)	Shin <i>et al</i> (2010)	Wei <i>et al</i> (2012)	Current Case
CD117 (c-Kit)	-	+ (epithelial cell)	+ (epithelial cell)	-
AE1/AE3	+ (epithelial cell)	+ (epithelial cell)	+ (epithelial cell)	+ (epithelial cell)
Chromogranin A	-	-	NA	-
Synaptophysin	-	-	NA	-
Vimentin	+ (spindle cell)	+ (spindle cell)	+ (spindle cell)	+ (spindle cell)
Desmin	-	-	-	-
S100	-	NA	-	-
CD34	-	-	NA	-
Calretinin	-	-	-	-
CD56	NA	+ (epithelial cell; focal in spindle cell)	+ (epithelial cell; focal in the spindle cell )	+ (epithelial cell and spindle cell)
CD10	+ (spindle cell)	+ (focal spindle cell)	+ (epithelial and spindle cell)	+ (epithelial and spindle cell)

Abbreviations: + positive; - negative; NA – not available

## **7. Differential diagnoses:**

- Biphasic synovial sarcoma
- Teratoma
- Carcinosarcoma

## **8. Take-home messages:**

- Gastroblastoma should be considered in the differential diagnosis of biphasic gastric tumours, especially in young patients.
- It represents a low grade malignant tumour
- Characteristic histological features such as mild atypia, low-mitotic activity and variable immunohistochemical staining with CD56 and CD10 are helpful for confirming the diagnosis.

## **V. REFERENCES:**

1. Miettinen M, Dow N, Lasota J, Sobin LH. A distinctive novel epitheliomesenchymal biphasic tumor of the stomach in young adults ("gastroblastoma"): a series of 3 cases. *Am J Surg Pathol.* 2009; 33:1370–1377.
2. Shin DH, Lee JH, Kang HJ, et al. Novel epitheliomesenchymal biphasic stomach tumor (gastroblastoma) in a 9-year-old: morphological, ultrastructural and immunohistochemical findings. *J Clin Pathol.* 2010; 63:270–274.
3. Wey EA, Britton AJ, Sferra JJ et al. Gastroblastoma in a 28-year-old man with nodal metastasis: proof of the malignant potential. *Arch Pathol Lab Med.* 2012; 136:961-964.