KARCINOSARKOMA E DUODENIT

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Summary

THIS REPORT DESCRIBES A CASE OF CARCINOSARCOMA OF THE DUODENUM

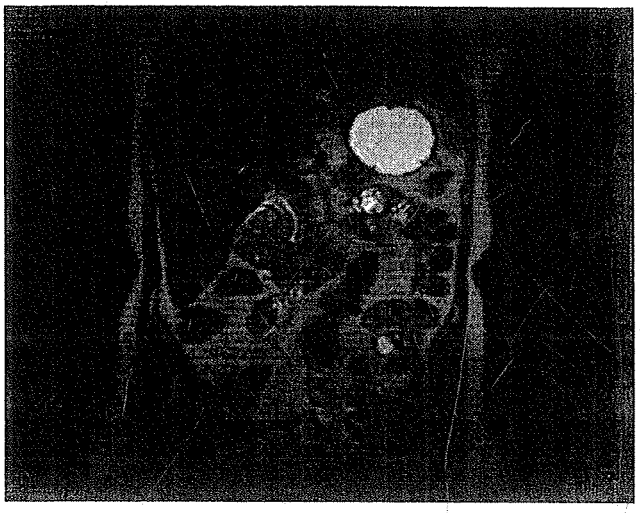
Carcinosarcomas are rare tumors, which include in their structure two histologic malignant components, one epithelial and one mesenchimal. They often are found in the head and neck region, respiratory tract and also in the reproductive female system. In the digestive system they are found more often in the orofarings or oesophagus but less often in the stomach. Carcinosarkomas rarely develop in the colon but when this happens they are extremely aggressive. Duodenal carcinosarcomas of the duodenum are rare, although this type of tumor is reported in other locations. We are presenting a case of Polypoid Carcinosarcoma of the duodenum in the Papilla Vater region in a patient of 59 years old, including the clinical, histopathologic and immunohistochemistry data. A polypoid tumor is located in the D2 portion of duodenum with a diameter of 7 cm resected together the pancreatic tissue from the head of the pancreas. The tumor was diagnosed as Popypoid carcinosarcoma of the duodenum with a wide peduncul infiltrating the Vater papilla. It contains two histologic components, one of well-differentiated adenocarcinoma that covers and infiltrates the tumor and a malignant fusiform component that serves as a stalk to the tumor. Immunohistochemistry examinations result in PanCK MNF positivity in the adenocarcinoma component, Vimentin positivity and Pan CK MNF negativity in the mesenchimal component, Ki67 very high in both of the components, neuroendocrine markers Synaptophisine and Cromogranin negative in the tumor cells, and hormonal receptorial status for Estrogen and Progesteron are negative.

Key words : Carcinosarcoma · Duodenum · Cefalopancreatoduodenectomy.

Raportim rasti

Karcinosarkomat jane tumore shume te rralle, qe perfshijne ne strukturen e tyre dy komponente histologjike malinje, ate epitelial si dhe ate mezenkimal [1]. Gjenden dy lloje te karcinosarkomes : karcinosarkoma e vertete dhe e ashtuquajtura karcinosarkome. Keto lloje klasifikohen ne baze te kontributit te karcinomes dhe sarkomes, kjo e lidhur ngushte me hipotezen e origjines monoklonale dhe multiklonale. Ky tumor eshte gjetur ne organe te ndryshme. Gjithsesi, karcinosarkoma e duodenit eshte shume e rralle. Vetem tre raste jane raportuar deri tani ne literature, dy nga te cilet pershkruajne zhvillimin e tumorit ne ampulen e Vater-it , rasti i trete raporton karcinosarkome ne duoden jashte ampules se Vater-it (2,3,4). Dhe rasti jone qe pershkruan karcinosarkome qe perfshin ampulen e Vater-it.

Pacientja A.M. 59 vjecare u paraqit prane klinikes se Gastrohepatologjise me nje histori 7 mujore per diskomfort epigastrik ,nauze,renie ne peshe dhe anemi. Laboratori rezultoi ne keto te dhena; Hb=8.0 g/dl, RBC=3,34x10^6/mm^3. Ekzaminimi endoskopik zbuloi nje mase okluzive ne porcionin D2 te duodenit. Me pas pacientes iu krye rezonanca manjetike, e cila zbuloi nje mase te pedunkuluar qe perfshinte ampulen e Vaterit dhe mbyllte lumenin e duodenit, pa zgjerime te pemes biljare apo duktusit pankreatik dhe pa perfshirje te limfonodujve regjional (Figura nr. 1/a,b).

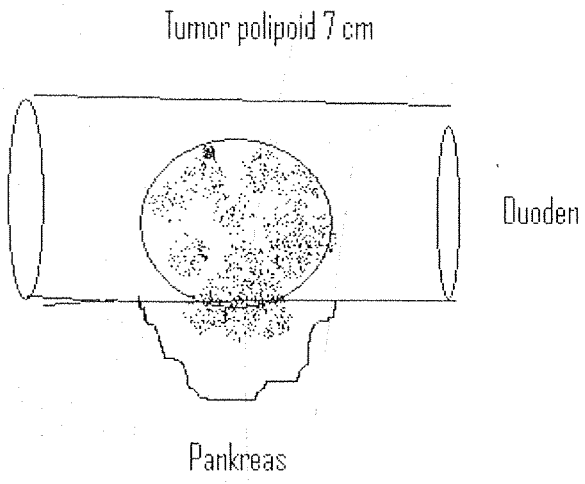


a

b

Figure nr.1 (a,b) MRI zbuloi nje mase te pedunkuluar qe perfshinte ampulen e Vaterit dhe mbyllte lumenin e duodenit.

Pacientja drejtohet ne repartin e kirurgjise ku kryen cefaloduodenopankreatektomi sipas Whipple me diseksion te limfonodujve. Materiali i perftuar pas kirurgjise tregon makroskopikisht nje tumor polipoid ne nivelin e D2 me diameter rreth 7 cm bashke me me nje pjese te kokes se pankreasit. (Skema) (Figura nr.2).



Skema

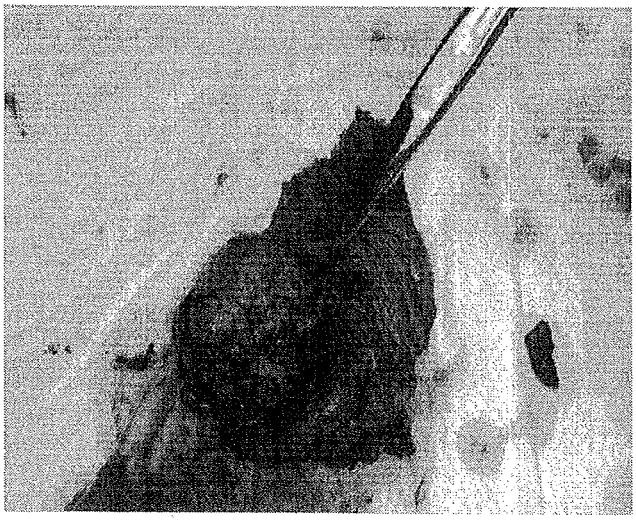


Figure nr.2 Materiali i marre pas kirurgjise

Klasifikimi TNM i tumorit u konkludua si $T_4N_0M_X$.

Histopatologjia-Tumori u diagnostikua si: E ashtuquajtur karcinosarkome polipoze me pedunkul te gjere dhe infiltrim te papiles se Vater-it qe permban dy komponente histologjike: Adenocarcinome te mirediferencuar mbuluese dhe infiltruese si dhe nje komponent fuziform malinj qe sherben si bosht mbeshtetes i formacionit. Tumori paraqet infiltrim te murit te duodenit pertej muskulatures propria. Kufinjte e rezeksionit jane te paster si dhe nuk u gjeten limfonodula metastatike ne 4 limfonoduj te ekzaminuar.

Imunohistokimia -U realizua per komponentet *epitelial* PanCKMNF, e cila doli pozitive ne komponentin adenocarcinomatoz dhe Vimentin e cila doli negative. Per komponentin *mezenkimal* u realizua vimentin, e cila doli pozitive dhe PanCKMNF, e cila doli negative duke konfirmuar prezencen dhe te ketij komponenti. U realizuan analiza te karakterit neuroendokrin synaptophysin, kromogranin, te cilat dolen pozitive ne indin pankreatik dhe qelizat endokrine te mukozes normale por negative ne qelizat tumorale epiteliale dhe mezenkimale. Gjithashtu dhe per indeksin proliferativ u krye Ki67, e cila rezultoi shume e larte ne te dy komponentet. Persa i perket sensibilitetit hormonal ndaj ER dhe PgR, HER2 rezultuan negative, keshtu qe pacientja nuk perfiton mjekim me antiestrogjenik si dhe immunoterapi me trastuzumab.

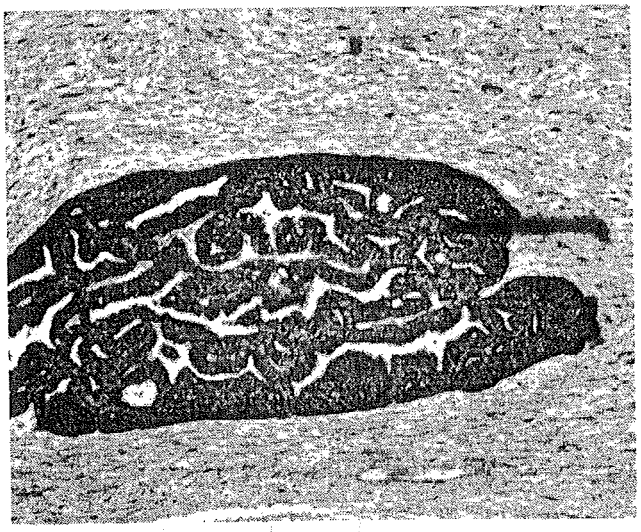


Figure nr.3 PanCK-MNF

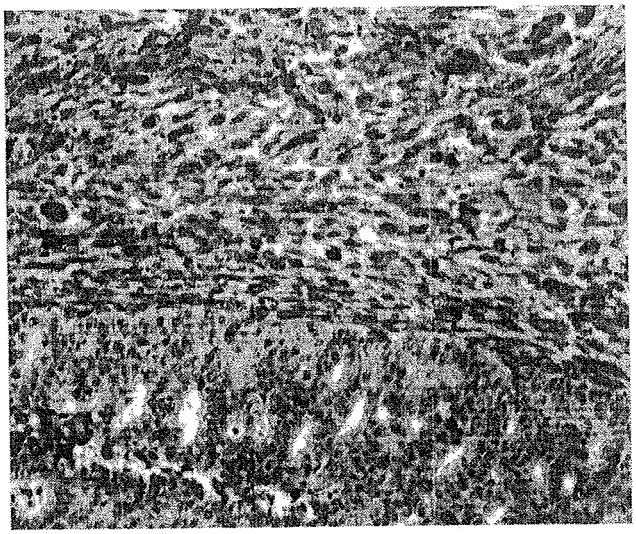


Figure nr.4 Vimentin

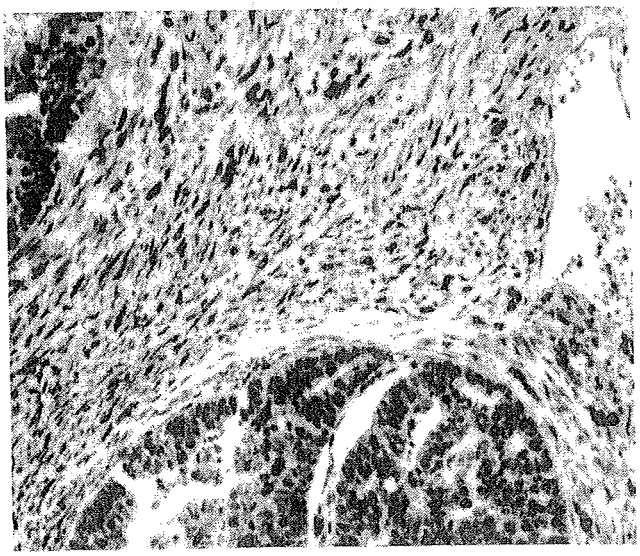


Figure nr.5 Ki67

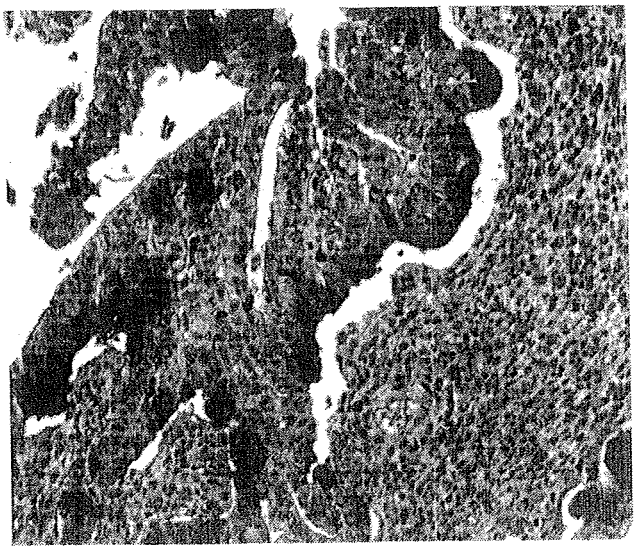


Figure nr.6 Cromogranin

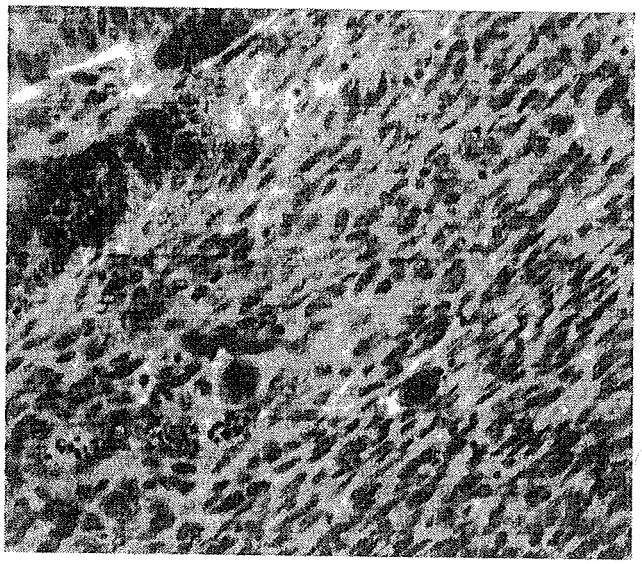


Figure nr.7 Synaptophisine

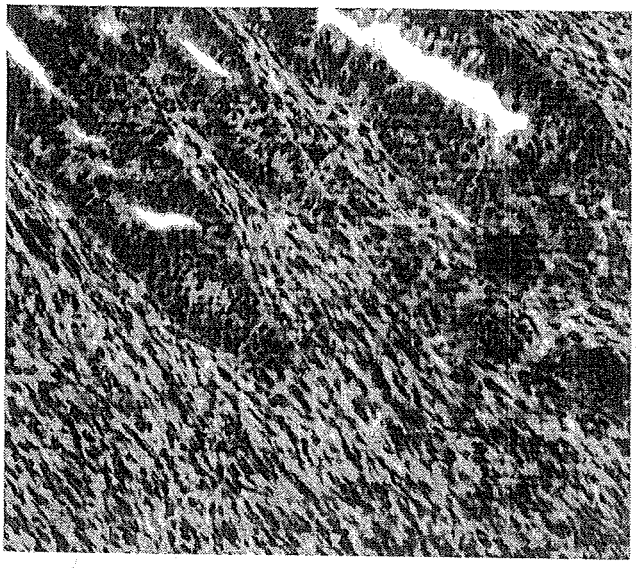


Figure nr. 8 Progesteron

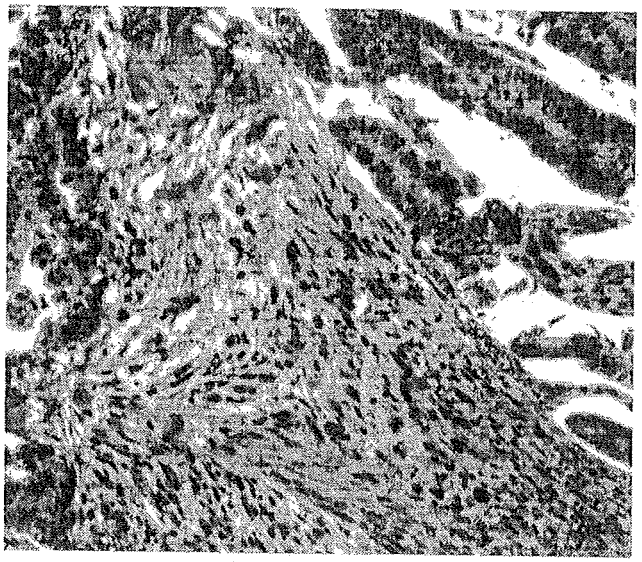


Figure nr.9 Estrogen

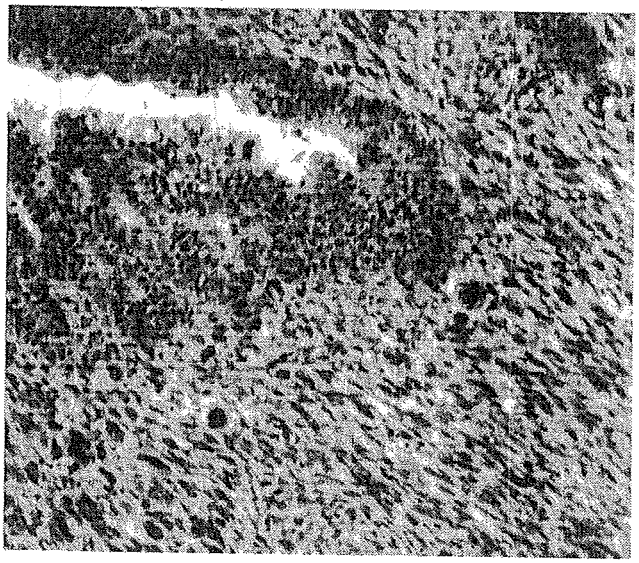


Figure nr.10 HER2

Pas nderhyrjes kirurgjikale dhe ekzaminimit imunohistokimik pacientes iu aplikua kimioterapi me cisplatin dhe adriamicine, 5 cikle. Pacietja doli ne gjendje te permiresuar nga spitali, dhe ne vazhdim kryen kontrolle te rregullta rutine.

Diskutim

Shpeshtesia e karcinomes se duodenit raportohet 0.02%-0.25% ne studimet autoptike (5). Shumica e tyre jane adenocarcinoma te mire dhe mesatarisht te diferencuara. Karcinosarkoma konsiston ne perzierjen e dy komponenteve: karcinomatoz dhe Sarkomatoz. Ishte Virchow-i ne 1864 i cili e raportoi si sarcoma carcinoides (6) ndersa Meyer unifikoi elementet nen termin e karcinosarcomes (7). Ato me shpesh shfaqen ne koke dhe ne qafe, ne traktin respirator, dhe ne organet ripodhuese femerore (8,9,10). Ne traktin gastrointestinal ato gjenden me shpesh ne orofarings, ezofag dhe me pak ne stomak. Karcinosarkomat rralle e marrin origjinen prej kolonit, por kur ndodhin ato jane ekstremisht agresive.

Kerkimi nepermjet PUBMED tregoi vetem tre raste ne literaturen e shkruar anglosaksone.

Keto raste paraqisnin nje grua dhe dy burra me moshe mesatare 54.6 vjec, ndersa rasti qe po raportojme behet fjale per grua 59 vjec (Tabela nr.1) (2,3,4).

Tabela nr.1 Raste te raportuara me karcinosarkome te duodenit

Autori	Mosha	Gjinia	Komponentet	Lokalizimi	Trajtimi	Prognoza
Kench 1997	46	F	Qeliza adenokarcinomatoze qeliza sarkomatoze (e ashtuquajtur)	Ne ampule	Pancreato-duodenectomy	Vdekur
Kijima 1999	46	M	Qeliza adenokarcinomatoze qeliza sarkomatoze (e ashtuquajtur)	Ne ampule	Pancreato-duodenectomy	Nukdihet
Sunagawa 2009	72	M	Qeliza adenokarcinomatoze qeliza sarkomatoze (e vertete)	Jashte ampules	Pancreato-duodenectomy	Gjalle
Rastiyne 2010	59	F	Qeliza adenokarcinomatoze qeliza sarkomatoze (e ashtuquajtur)	Ne ampule	Whipple	Gjalle

Tre jane teorite kryesore te histogjenezes se karcinosarkomave (11):

Teoria e pare paraqet komponentin sarkomatoz fuziform si zhvillim reaktiv nga karcinoma. **Teoria e dyte** konsideron si zhvillim te pavarur dhe te njekohshem te komponenteve malinje qe japin zhvillimin e karcinosarkomave te verteta.

Teoria e trete i konsideron elementet e vecanta te derivuara nga nje qelize meme e vetme (te ashtuquajturat karcinosarkoma). Ne pergjithesi karcinosarkomat e verteta kane origjine multiclonale (11), ndersa ato te ashtuquajturat karcinosarkoma kane origjine monoklonale (12). Prania e imunoreaktivitetit te citokeratines ne te dy perberesit, si ate karcinomatoz dhe ate sarkomatoz, sikurse ne rastin tone, mbeshtet origjinen epiteliale te perberesit sarkomatoz (13). Studimet me mikroskopi elektronike te qelizave sarkomatoide ne karcinosarkomat/karcinomat sarkomatoide kane treguar nje perzierje te tipareve te qelizave me karakteristika epiteliale (dmth lidhjet nderqelizore, tonofibrilet etj.) tek qeliza te tjera qe shfaqin karakteristika mezenkimale (14). Per me teper, sjellja biologjike e ketyre demtimeve duket me shume e ngjashme me Karcinomat "high grade" ne stade dhe vende analoge, sesa me sarkomat puro analoge qe shfaqen ne te njejtat organe. Prandaj disa autore propozojne qe keto tumore te quhen karcinoma sarkomatoide sesa karcinosarkoma (15). Megjithate ka disa raportime se ekzistojne shprehje te Cytokeratines dhe tek sarkomat puro te tipeve te ndryshme (16).

Perfundime

Shumica e karcinosarkomave te traktit tretes kane nje komponent qelizor fuziform. Ato jane prezantuar ne forma te ndryshme por shumica e tyre jane te ashtuquajtura karcinosarkoma. Gjithsesi histogjeneza e tyre mbetet per tu studiuar me tej. Karcinosarkomat e raportuara ne traktin tretes shoqerohen me prognoze te keqe. Gjithsesi, ato raste qe kane marre trajtim kurativ kane treguar mbijetese afatgjate.

BIBLIOGRAFIA

1. **Wick MR, Swanson PE.** Carcinosarcoma: current perspectives and an historical review of nosological concepts. Semin Diagn Pathol 1993;10:118–27.
2. **Kench JG, Frommer DJ.** Sarcomatoid carcinoma of the ampulla of Vater. Pathology 1997;29:89–91.
3. **Kijima H, Takeshita T, Suzuki H, Tanahashi T, Suto A, Izumika H, et al.** Carcinosarcoma of the ampulla of Vater: a case report with immunohistochemical and ultrastructural studies. Am J Gastroenterol 1999;94:3055–9.
4. **H. Sunagawa et al.:** Carcinosarcoma of the Duodenum
5. **Manier J. Duodenal tumors. In: Haubrich WS, Schaffner F, Berk JE, editors.** Bockus gastroenterology. 5th ed. Philadelphia: Saunders; 1994. p. 875–81.
6. **Virchow R:** Die Krankhaften geschwulst. Vol. 2A. Berlin: Hirschwald; 1864.
7. **Meyer R.** Beitrag zur Verstandigung uber die Namengebung in der Geschwulstle hre. Zentralbl Allg Path Anat 1919;30: 291–6.
8. Iyomasa S, Kato H, Tachimori Y, Watanabe H, Yamaguchi H, Itabashi M. Carcinosarcoma of the esophagus: a twenty-case study. Jpn J Clin Oncol 1990;20:99–106.
9. **Nappi O, Glasner SD, Swanson PE, Wick MR.** Biphasic and monophasic sarcomatoid carcinomas of the lung. A reappraisal of "carcinosarcomas" and "spindle-cell carcinomas". Am J Clin Pathol 1994;102:331–40.
10. **Tavassoi FA.** Classification of metaplastic carcinomas of the breast. Pathol Ann 1992;27:89–119.
11. **Madan AK, Long AE, Weldon CB, Jaffe BM.** Esophageal carci- nosarcoma. J Gastrointest Surg 2001;54:414–7.
12. **Thompson L, Chang B, Barsky SH.** Monoclonal origins of malig- nant mixed tumors (carcinosarcomas). Evidence for a divergent histogenesis. Am J Surg Pathol 1996;20:277–85.
13. **Zarbo RJ, Crissman JD, Venkat H, Weiss MA.** Spindle cell carcinoma of the upper aerodigestive tract mucosa. An immunohistologic and ultrastructural study of 18 biphasic tumors and comparison with seven monophasic spindle cell tumors. Am J Surg Pathol 1986; 10:741-53.
14. **Battifora HI** Spindle-cell carcinoma: ultrastructural evidence of squamous origin and collagen production by the tumor cells. Cancer 1976; 37: 2275-82.
15. **Iezzoni JC, Mills SE,** Sarcomatoid carcinomas (carcinosarcomas) of the gastrointestinal tract: a review. Semin Diagn Pathol 1993; 10: 176-87.
16. **Lawson CW, Fisher C, Gatter KC.** An immunohistochemical study of differentiation in malignant fibrous histiocytoma. Histopathology 1987; 11: 375-83.