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Pigmented neuroendocrine carcinoma of the hepatic duct: a case report

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Background

Neuroendocrine tumors of the gallbladder or the extrahepatic bile ducts are rare. We describe a pigmented neuroendocrine carcinoma of the common hepatic duct with a regional lymphnode metastasis. We reviewed the literature for these tumors and discuss the nature of the pigment.

Methods

The clinical and macroscopic appearance was documented together with a preoperative cytology. Paraffin-embedded tissue was used for histological, histochemical and immunohistochemical analysis. An electron microscopy analysis was performed.

Results

A pigmented tumor measuring 2.9 cm was located in the common hepatic bile duct with lumen stenosis reaching to the bifurcation of the hepatic ducts. The preoperative cytology showed atypical and single pleomorphic tumor cells with pigment granules and an immunocytological positive reaction for cytokeratin. The histological examination revealed an unencapsulated neoplasm infiltrating the biliary wall including the mucosa. The neoplastic cells were organized in solid, trabecular structures or cords in a condensed stroma. They were argyrophil in the Grimelius and non-argentaffin in the Fontana-Masson silver impregnation, while the pigment granules were argentaffin. The tumor cells exhibited a diffuse strong staining for chromogranin A, neuron-specific-enolase, a lesser staining for synaptophysin and neurofilament, and a weak nuclear reaction for S-100 protein. Cytokeratin was positive and HMB45 completely negative. No S-100 positive sustentacular cells could be found. The proliferation-index Ki-67 (MIB1) was low (2%). The electron microscopy revealed electron-dense neuroendocrine granules.

Conclusion

Neuroendocrine tumors of the extrahepatic bile ducts represent only 0.1 to 0.2 percent of all neuroendocrine tumors of the gastrointestinal tract. In the English literature, nearly 40 cases are reported. Pigmented neuroendocrine tumors reported previously are mainly bronchopulmonary carcinoids. In our review of the literature, a pigmented neuroendocrine tumor was not described for the extrahepatic biliary ducts before. The pigment in these tumors is considered as melanin pigment, lipofuscin or neuromelanin. In some cases, the nature of the pigment could not be designated. Pigmented neuroendocrine tumors are an important differential diagnosis in pigmented tumors because the black pigment suggests the diagnosis of metastatic melanoma. In the present case, the preoperative documented immunocytological positivity for cytokeratin prevented the diagnosis of metastatic melanoma in the frozen section.