Chirurgia (Bucur). 2012;107(3):404-407

Obstructive jaundice by genodermatosis – case report

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Abstract

Background: Neurofibromatosis type I, Recklinnghausen disease is the most frequently of neurofibromatosis occurs in 1/3000-1/5000 of children born. Disease is a genodermatosis with 1/3000-1/5000 autosomal dominant transmission. Incriminated in the pathological appearance of the disease gene is located on chromosome 17, gene product, neuro-fibromina, is a protein involved in controlling cell differentiation and proliferation. Skin manifestations can be associated with the same papillary tumors and the internal organ. Treatment is surgery for larger tumors. Worse prognosis in malignant developpment, with the lower quality of life in the presence of complications, as in this case: mechanical obstructive jaundice.

Material and method: Patients aged 75 years, admitted for obstructive jaundice (progressive, pruritic), cutaneous papillomas (0.5-3 cm) on the trunk and several hyperpigmented spots, brown (5-6 cm diameter). Cutaneous lesions (45 years old) have been previously diagnosed by histological examination.

Results: Surgery, under general anesthesia we did: cholecystectomy, intraoperative choledocoscopy of bile duct. In the last portion of bile duct we founded pediclled tumors. We did partial excision of tumors and coledoco-duodenoanastomosis in healthy tissue. Histological examination shows neuro-fibrodermatoza type 1. Discharge 12 days postoperatively after.

Conclusions: Preoperative diagnosis suggested the possibility of mechanical jaundice by malignancy. Etiologic diagnosis of this rare form of obstructive jaundice could not be established before surgery, only by histological examination of tumors removed.

Key words: neurofibromatosis, obstructive jaundice, papillomas, coledoco-duodenoanastomosis

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