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An astonishing familiar cluster of Intrahepatic Cholangiocarcinoma (ICC) in Wilson Disease (WD)

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Introduction : According to literature, the rate of hepatobiliary malignancies in WD is low; occasional cases of hepatocellular carcinoma (HCC) have been reported with even rarer incidence of ICC. We report the case of a family with WD in which two siblings, Mr F and Mr G, developed ICC at the ages of 56 and 60 respectively while their sister was affected by WD without primary liver malignancies.

Methods : We made a literature search for publications including age at ICC diagnosis, time between diagnosis of WD and tumor detection and type of therapy.

Results : Literature data report up to now only thirteen cases of ICC associated with WD. Nothwithstanding considerable heterogeneity of treatment all the patients who underwent surgical resection were alive after two years. Mr F underwent primary surgery for ICC and he relapsed after one year. The patient, therefore, had liver transplantation in November 2018 after conversion chemotherapy with Gemcitabine - Cisplatin schedule achieving a surprising complete response. Next Generation Sequencing (NGS) found an undescribed FGFR2 oncogenic somatic mutation. Mr G developed ICC and HCC one year later. The two malignancies were treated with surgical resection and the patient is currently without any evidence of recurrence of the disease after four years.

Conclusions : While the reasons for the low incidence of ICC in WD remains obscure, we underline the exceptional findings of two ICC in two siblings in absence of additional definite risk factors, one of which showed complete response to chemotherapy.

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