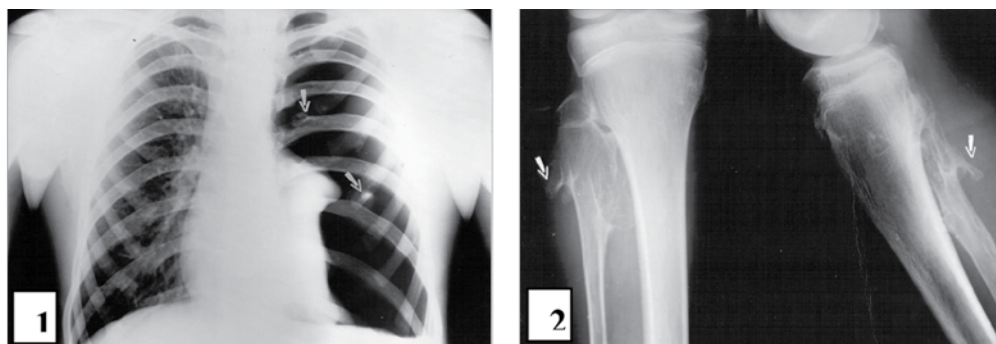


PNEUMOTHORAX AND PERONEUS NERVE LESIONS IN THE BOY WITH MULTIPLE FAMILIAR EXOSTOSIS

Vojko ROŽMANIĆ, Neven FRLETA

Pediatric Clinic, Clinical Hospital Centre Rijeka, University of Rijeka, Croatia
e-mail: vojko.rozmanic@medri.hr; tel.: + 385 51 659 111



Multiple hereditary exostoses (MHE) are heterogeneous, autosomal dominant disorders of mostly the long bones, characterized by formation of benign bony and cartilage tumors in multiple locations. Multiple hereditary exostoses are rare. Their incidence is 1:50,000. Exostoses are usually localized on preference areas, primary at the juxtaphysal regions of long bones, however, they may also be formed on other parts of the skeleton. This means that, within MHE, we can find benign cartilage and bony tumors even on the pelvis, shoulder blade, ribs, heel bone and vertebrae. They never appear on the face or skull bones. Symptoms are mainly connected to the tumor localization. It is not recommended to remove asymptomatic exostoses. In those cases there is great possibility of relapse (20-50%). Exostoses that deform the esthetic, rather than the functional integrity of a patient are indicated for extirpation with the possibility of relapse. It is recommended that formations that are threatened by their localization, bear the risk of the latent development of severe complications and therefore should be removed. A rare, but severe complication is a malign alteration, occurring in 1% of patients suffering from MHE, but generally in adults. A 16 year old boy with a negative family history developed left side pneumothorax. Multiple exostoses on the 4th and 5th ribs were noticed. Following thoracocentesis complete lung relapse was accomplished. No further complications were noticed. However, 6 months later, symptoms of a lesion on the left peroneal nerve occurred. After comprehensive neuropediatric management, including EMG, paralysis of the n. peroneus was determined, with palpable exostoses in the region of the proximal left tibia. Two metaphase exostoses of the left fibula were removed and decompression of the left peroneal nerve achieved. The postoperative course was satisfactory with regular physiotherapy. After full management we could easily conclude that compression of the two exostoses in the nerve canal led to the paralysis of the peroneal nerve, which was confirmed during surgery, and after decompression it slowly led to complete regression of the symptoms. On the other hand, pneumothorax is a very rare complication in MHE patients. X-ray management confirmed the presence of exostoses on the ribs, which could have led to the development of this condition. However, spontaneous pneumothorax cannot be excluded.