A giant chondrosarcoma of the anterior chest wall together with multiple hereditary exostoses of the lower extremities is presented. The 20-year-old male patient had been living with the lesion for 14 years during which time it had grown to giant dimensions (Fig. 1). The tumour was excised en bloc and the defect was repaired using just skin flaps. The pedigree tree is shown in Fig. 2.

**Fig. 1.** The giant mass lesion (chondrosarcoma) of $20 \times 18 \times 20$ cm dimensions originating from the right arcus costarium together with bilateral multiple exostoses at the lower extremities are seen.

**Fig. 2.** Pedigree tree. Individual IV 6, who transmitted the gene to his children, died of leukemia. Proband is the only one who had chondrosarcoma. The patient’s chromosome analysis revealed normal karyotype with polymorphism of 46, XY, Inv 9 (p1.1, q1.3). These could have been an example of EXT 1 mutation and LOH on this locus.