Current Problem Cases

Familial Multiple Neurilemoma

A Case Report

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Summary. A case of familial multiple neurilemoma is reported. A 59-year-old man with five neurilemomas was treated. Fifteen of his 22 family members had had soft-tissue tumors. Ten neurilemomas had been diagnosed microscopically in five members of the family. There were no symptoms related to von Recklinghausen’s disease. This case seems to be a forme fruste or related form of von Recklinghausen’s disease.

Zusammenfassung. Es wird ein Patient mit familial vorkommenden multiplen Neurilemmomen vorgestellt. Es handelt sich um einen 59 Jahre alten Mann, bei dem insgesamt 5 Neurilemmome chirurgisch entfernt wurden. In der Familie des Patienten hatten 15 von 22 Mitgliedern Weichteiltumoren, wovon bei 5 Familienmitgliedern insgesamt 10 Neurilemmome mikroskopisch diagnostiziert wurden. Obwohl es sich bei dem vorgestellten Fall nicht um die klassische Form eines Morbus Recklinghausen handelt, vermuten die Autoren doch einen gewissen Zusammenhang oder das Vorliegen einer Form Fruste.

Benign neurilemoma usually occurs as an encapsulated solitary neoplasm arising from Schwann cells, which grows in an expansile manner. Although it arises in patients who have no evidence of a genetic predetermination, it is occasionally a component of neurofibromatosis (von Recklinghausen’s disease).

In this paper we present a case of familial multiple neurilemoma inherited as an autosomal dominant trait, unassociated with other symptoms such as café-au-lait spots, neurofibromatosis of the skin, or bone deformities which occur in von Recklinghausen’s disease.

Case Report

A 59-year-old man was referred to us in September 1985 with an 8-year history of a painful tumor in the region of his right elbow. The tumor had grown year after year, and he also complained of constant sciatica, especially at dawn. He had already undergone surgery for three neurilemomas in our hospital. On his first admission, at the age of 32, a neurilemoma of the right median nerve had been excised. Then, at the ages of 44 and 51 respectively, two neurilemomas of the right femoral nerve and the left sciatic nerve had been excised.

The pedigree of his family members is shown in Fig. 1. There was a markedly frequent occurrence of soft-tissue tumors in his family. His father and uncle were said to have had some soft-tissue tumors but had not undergone surgery. His five brothers and a sister had also had some tumors. Four of these that had been removed and examined microscopically were neurilemomas. His son and two nieces had had multiple soft-tissue tumors, for which the latter had undergone surgery at another hospital, and the pathological diagnosis was also neurilemoma. Ten neurilemomas had been diagnosed microscopically in five members of this family.

There were no symptoms related to von Recklinghausen’s disease, such as café-au-lait spots, neurofibromatosis, bone deformities, or ocular lesions in the patient and his family members. No history of malignancy or consanguineous marriage was known among them.

Examination of the right elbow revealed a 3 x 3 cm elastic, soft, tender tumor at the scar from a previous operation. There was no muscle weakness and no alteration in skin sensation of
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Fig. 1. Pedigree of family members. Fifteen of 22 had had soft-tissue tumors. Ten tumors in five members were removed and confirmed to be neurilemmas.

Fig. 2. Myelograms show an undulated piece of cauda equina leading to a small round defect at left L4–5 (arrows).

Fig. 3. Histological features of the median nerve tumor: a typical neurilemoma, Antoni type B.

The right upper limb. A slight sciatic scoliosis was noted in the lumbar spine. The left SLR test was positive. The left ankle jerk was absent. There was marked impairment of power of all peroneal muscles, and skin sensation was diminished at the lateral aspect of the thigh and the medial aspect of the leg. Radiographs of the spine showed no abnormalities or deformities except for mild degenerative changes. However, metrizamide myelograms demonstrated an undulated piece of cauda equina leading to a small round filling defect at the left L4–5 disc space (Fig. 2). The diagnosis of a cauda equina tumor was made on the basis of these findings.

In September 1985, the tumor of the right elbow was removed. It was 3 x 3 cm, with a fibrous capsule on the median nerve trunk, and two funiculi were riding on the tumor. The tumor was enucleated, with preservation of its parent nerve. Histological examination of the tumor showed a typical neurilemoma, Antoni type B (Fig. 3). In November 1985, laminectomy from L4 to S1 was done, and the cauda equina tumor was