Letters to Editor

Nuchal extra-abdominal aggressive fibromatosis of desmoid type in a 77-year-old female

Sir,

A 77-year-old female noticed a fast growing, indolent lesion within the left nuchal musculature. Ten years ago, she was operated for renal cancer. Preoperative MRI showed a sharply demarcated lesion [Figure 1A] with a strong uptake of gadolinium [Figure 1B] after administration of 0.4mmol gadolinium-DTPA per kg body weight. X-ray of the cervical region displayed the tumor [Figure 1C]. Because of aggressive growth, we supposed a metastasis of the renal cancer and immediately we performed radical tumor excision.

The tumor exhibited a high cellularity [Figure 2] with monomorph spindle cells in herringbone arrangement between dense collagenous fibers. Pycnotic nuclei indicated apoptosis [Figure 2]. Maximally three mitoses were counted within 50 high-power fields. A formation of fissures was exhibited around blood vessels. Small perivascular cuffs of lymphocytes completed the...
microscopic picture. There was a tendency to infiltrate the peritumoral muscle, which showed degeneratively changed giant cells. The diagnosis of a nuchal extra-abdominal aggressive fibromatosis of desmoid type was established.

Aggressive fibromatosis is a rare tumor of the soft tissue with an incidence of two to four per million. Normally neurosurgeons are seldom confronted with this entity.\(^1\)

In general it is a tumor of younger patients. Mean age at presentation is 30 to 45 years.\(^2,3\) Our 77-year-old patient is extraordinarily old.

Because of its destructive growth into the surroundings the tumor is semimalignant.\(^2,4\) Distant spread hasn’t been observed yet, but significant mortality can result from local tumor complications. Total tumor resection within healthy tissue is therefore the most important prognostic factor, otherwise the rate of local recurrences is high.\(^1,2\)

Clinically, patients suffer from an indolent mass relatively fast growing sometimes causing symptoms by compressing adjacent structures, e.g. the plexus brachialis.\(^1\) The shoulder, upper arm and the gluteal region is the main location in children. Our patient presented the typical history, but the location in the neck was unusual. Mostly females are affected with a percentage of about 66% of all described cases.\(^2\)

Magnetic resonance imaging is the method of choice to visualize the tumor with its borders. Conventional X-ray of the region often shows a diffuse soft tissue swelling. Possible differential diagnoses are inflammatory changes such as fasciitis nodularis and other lesions such as fibromas, leiomyosarcomas, myopericytommas, myofibroblastomas, chondroblastomas, metastatic spindle cell carcinoma of the breast and myomas.\(^3\) But the predominant fibrous differentiation or a differentiation along smooth muscle was not observed in our patient’s tumor material. Some describe aggressive fibromatosis to be a lesion that imposes with homogenous isointensity or mild hyperintensity in T1-weighted sequences of MRI.\(^3\) In our case there was a mild hyperintensity on T1-sequences [Figure 1A]. A heterogeneous high signal was seen in T2-sequences [Figure 1C]. All the lesions published enhanced inhomogenously gadolinium after intravenous administration.\(^3\) The homogenous enhancement of our patient’s tumor could be related to a high dosage of gadolinium or the very high cellularity of the lesion. Another prominent difference was the appearance of the tumor as a discrete mass in comparison to an infiltrative ill-defined enlargement of a muscle group in other cases.\(^3\)

Gross total resection within healthy tissue is the treatment of choice. It is the only well-known prognosis parameter.\(^2,3,5\) Incomplete resection seems to be associated with a higher rate of recurrence. The overall recurrence rate is 35-60%. Patients with complete tumor resection had a significantly longer recurrence-free interval than patients in whom resection was incomplete with 83 versus 60% five-year relapse-free interval. Adjuvant radiotherapy seems to have only little effect on recurrence and chemotherapy has been ineffective up to now.\(^2,5\) Spear et al found local control rates of 77% with radical surgery alone versus 100% for patients with surgery and radiation after microscopically complete resection.\(^6\) So, the overall prognosis depends mainly on the localization of the process.

Neurosurgeons are seldom confronted with extraperitoneal aggressive fibromatosis of the desmoid type. It is necessary to know the essentials of this neoplasm in order to manage it correctly. Complete surgical removal still remains the treatment of choice; whereas radiation therapy is reserved for situations in which gross resection is not possible because of cosmetic or functional reasons.

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References

Accepted on 23-05-2007