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Epithelioid schwannoma of the ulnar nerve

Some clinical observations

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The authors describe one case of purely epithelioid schwannoma of the ulnar nerve and discuss the therapeutic management.
A 44 year-old man was referred to us for a fusiform, movable mass in the left ulnar nerve and was removed by a wide en bloc excision. Intraoperative nerve action potentials were performed both prior to and following excision of the lesion. Histologically, the tumor was composed of round or polygonal cells arranged in necrotic clusters and anastomosing cords. There were areas of spindle cells. The epithelioid cells were round with abundant cytoplasm. Mitoses were frequent. S100 protein immunoreactivity was present diffusely in tumor cells (both nuclear and cytoplasmic), whereas cytokeratin, NSE, and anti-melanoma reactions gave negative results. After a 13 months, the patient's neurological conditions are excellent and there are no signs of either recurrence or metastasis.

Key words: **Peripheral nerve neoplasms - Ulnar nerve - Neurilemmoma surgery.**

MPNST (malignant nerve sheath tumors) are tumors of neuroectodermal origin that account for about 10% of all soft-tissue sarcomas and their incidence among nerve sheath tumors ranges from 2% to 12.7%.[1-4] Most published reports of epithelioid MPNST consist of only a few cases,[3-9] but the frequency of the "purely epithelioid" MPNST is extremely rare.[1 2 5 10 11]

We describe one case of purely epithelioid MPNST and examine their therapeutic and prognostic features.

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Case report

A 44 year-old man with no stigmata of NF was referred to us for a 2-year history of a tender mass behind his left arm and noticed that the movement of this arm was accompanied by local pain that had begun 3 months earlier. On neurological examination he presented a painful mass in the left arm.

CT showed a dyshomogeneous lesion at the superficial level of the left arm. MRI showed a dyshomogenous superficial lesion, with peripheral enhancement after gadolinium, arising from the ulnar nerve (Fig. 1 A, B).

The patient was submitted to surgery: a fusiform, brown mass of the left ulnar nerve in the left arm. The fusiform mass was movable within the surrounding connective tissues and was removed by a wide en bloc excision. Intraoperative nerve action potentials were performed both prior to and following excision of the lesion to identify viable

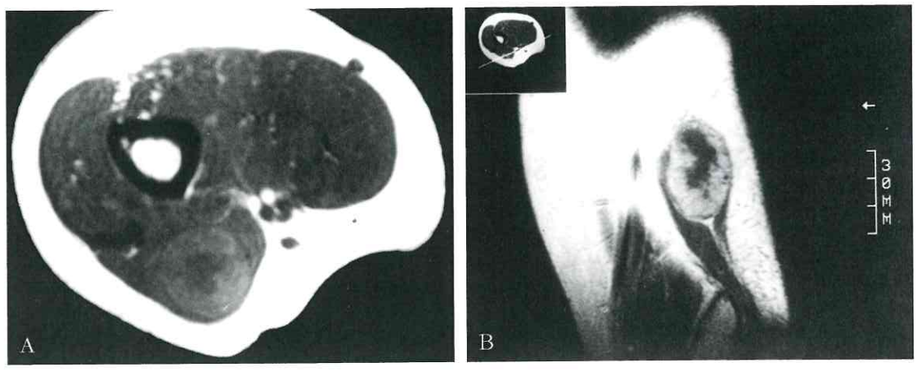

Fig. 1.—MRI axial (A) and coronal (B) views. T1-weighted image showed a dyshomogeneous lesion arising from the ulnar nerve (A) with peripheral enhancement (B).

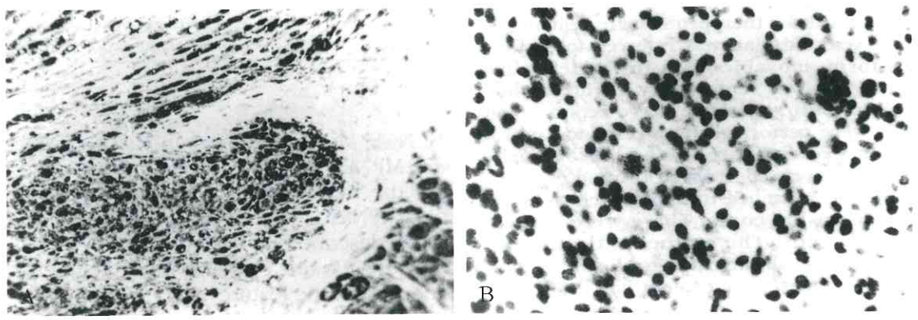

Fig. 2.—Neoplastic cells with vescicular nuclei and eosinophilic cytoplasm (A) (H & E, ˜450); cells showing cytoplasmatic positivity for S100 protein (B).

nerves, an attempt was made to resect as much of the tumor as possible while preserving normal nerve fibers. The tissue removed was small (3 cm) and consisted of fusiform mass, lobulated, variably pink, pale-tan to brown. Histologically, the tumor was composed of round or polygonal cells arranged in necrotic clusters and anastomosing cords. There were areas of spindle cells. The epithelioid cells were round with abundant cytoplasm (Fig. 2 A, B). The nuclei were vesicular and contained nucleoli. The cytoplasm was generally abundant and eosinophilic, with distinct borders. Mitoses were frequent (3 of 10 in high-power field). S100 protein immunoreactivity was present diffusely in tumor cells (both nuclear and cytoplasmic), whereas cytokeratin, NSE, and anti-melanoma reactions gave negative results.

Postoperative course was uneventful and the patient left hospital without any changes in his preoperative neurological deficits. Thirteen months later he did not present any clinical or radiological (MRI) signs of recurrence.

Discussion and conclusions

About one-half of patients with peripheral malignant schwannoma have evidence of neurofibromatosis.[1 5] Epithelioid MPNST seem to have a lower association with NF; none of the 26 patients reported by Laskin *et al.*[3] had NF and only one of the 14 patients reported by Lodding *et al.*[4] was so affected.

Purely epithelioid schwannoma can easily be mistaken for other tumor types. Prominent among these are metastatic carcinoma, sweat gland carcinoma, and malignant melanoma.[11-14] The most reliable evidence in support of a diagnosis of epithelioid MPNST is the demonstration of origin from a nerve, as in our case. Positivity for S100 protein and negativity for cytokeratin and anti-melanoma antiserum is very helpful also.

The sex (males) and age distribution (IV decade of life; range 5-80 years) corresponds to benign schwannomas.[15] The majority of those tumors originated in major nerves (including the sciatic, tibial, peroneal, facial, antebrachial cutaneous and digital); however, origin from a nerve cannot always be documented.[3 4 7 16] Clinical data, CT scan and MRI pictures do not permit differentiating malignant from benign schwannomas, except when an ill-defined tumor infiltrates soft tissue.[2 16 17] CT is sensitive to the cystic change that frequently accompanies these tumors and it has also been praised as the diagnostic test of choice for tumors of the sciatic nerve. MRI is capable of reliably imaging not only the tumor and its capsule, but also the nerve from which the tumor arises. In our cases, CT scan and MR images did not permit a differential diagnosis.

The appropriate management of this tumor is not well established. Radical surgical excision (resection as much of the tumor as possible preserving normal nerve fibers) has been reported as the primary modality of treatment, whereas the role of adjuvant radiotherapy and chemotherapy appears unsettled.[8 14 18 19] Incomplete removal of tumor caused the worse prognosis (recurrence and metastases), but recurrence is possible after total removal as well.[5] Di Carlo *et al.*[6] in a review of 8 "purely epithelioid" MPNST noted that 5 patients were alive up to 2.5 years later and 3 were alive and well up to 4 years later. Laskin *et al.*[3] noted that only one of 16 patients with superficial tumor developed a distant metastasis; instead, three of 10 patients with deep tumor died of metastatic disease. The author concluded that superficial epithelioid MPNST has a favourable course and must be considered a "limited stage" lesion. Perhaps, as in our case, cure could be obtained with radical surgery alone when the tumor is small. In fact, microscopic tumoral diffusion in contiguous tissues, responsible for development after total tumor removal, can reasonably be expected in larger tumors.[2 11 16 17 20]

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Riassunto

Neurinoma epitelioide del nervo ulnare. Osservazioni cliniche.

Gli Autori descrivono un caso di neurinoma epitelioide «puro» del nervo ulnare e ne discutono il trattamento.

Un uomo di 44 anni è giunto alla nostra osservazione per la presenza di una massa fusiforme e mobile del nervo ulnare sinistro ed è stato rimosso in blocco. Durante l'intervento, prima e dopo la rimozione della lesione, sono stati eseguiti i potenziali d'azione del nervo. Istologicamente, il tumore era composto da cellule rotondeggianti o poligonali organizzate in grappoli con necrosi ed in cordoni interlacciati. C'erano aree di cellulefusate. Le cellule epiteliodi erano rotonde con abbondante citoplasma. Le mitosi erano frequenti. Nelle cellule tumorali era presente una immunoreattività difesa (sia nucleare che citoplasmatica per la proteina S100), mentre l'immunoreattività per la citocheratina, la NSE e anti-melanoma erano negative. Dopo 13 mesi, le condizioni neurologiche del paziente sono eccellenti senza segni neurologici di recidive o metastasi.

Parole chiave: Neurinomi maligni - Neurinoma epitelioide - Sistema nervoso periferico, neoplasie - Nervo ulnare.

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Anastomosi intestinali con anello bioframmentabile (BAR)

Risultati e confronto con le anastomosi meccaniche e manuali

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Large bowel anastomosis with biofragmentable ring (BAR). Results and comparison with stapled and conventional suture.

Methods. **The use of the biofragmentable anastomosis ring (Valtrac-BAR) was attemped in 25 selected patients (group I) undergoing elective colonic resection for primary colon cancer, at the General and Cardiovascular Institute of the University of Milan. The results were compared with those of 30 selected patients who underwent elective colonic resection for the same pathology during the same period, and had their bowel anastomosis stapled (group II) or hand-sutured (group III).**
Results. **In group I there were 2 complications requiring reoperation: a little tear of the bowel near the BAR, and a postoperative ileus, compared with none in group II, that developed only 3 cases of abdominal wall infections. In group III, 2 patients developed a leakage of the anastomosis, one of them required reoperation. In the perioperative course there were 2 deaths (3.6%), in group II and III due to cardiovascular diseases. The results obtained showed no significant difference in the clinical course of the patients, or time of return of bowel function, and hospital stay.**
Conclusions. **The conclusions is drawn that Valtrac-BAR device is a safe and reliable alternative to conventional suture anastomosis in all areas of the intestinal tract, except the low rectum.**

Key words: **Valtrac-BAR - Colorectal neoplasms surgery - Anastomosis, surgical instrumentation - Surgical stapling - Anastomosis, surgical methods.**

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La possibilità di realizzare un'anastomosi intestinale senza l'impiego di materiali di sutura è stato per molti anni uno degli argomenti più discussi ed indagati nell'ambito della chirurgia gastroenterica. Fin dal 19° secolo, i chirurghi dell'epoca avevano ideato diversi e spesso complicati dispositivi per la realizzazione di anastomosi intestinali di questo tipo. Nel 1826, Denans [1] dopo le fondamentali sperimentazioni di Lembert [2] sulle tecniche di sutura introflettente nella realizzazione delle anastomosi intestinali, inventò un dispositivo cilindrico d'argento costituito da tre anelli ad incastro che mantenevano affrontati per compressione a margini introflettenti i 2 monconi intestinali fino alla loro sintesi biologica, mentre la compromissione della circolazione locale nei segmenti intestinali così affrontati, ne causava la necrosi locale con