Monophasic synovial sarcoma of hypopharynx: case report and review of the literature

Sarcoma sinoviale monofasico dell'ipofaringe: descrizione di un caso clinico e revisione della letteratura

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Parole chiave

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Summary

Synovial sarcoma (SS) is a malignant mesenchymal neoplasm usually involving the lower limbs of young adults. Localization in head-neck district is rare. Histologically, these are characterised by a biphasic or monophasic variant, the latter being more rare and difficult to identify. Immunohistochemistry plays a crucial role in the diagnosis. Cytogenetics also play an important role since both the monophasic and the biphasic forms are characterised by a reciprocal translocation (x;18) (p 11.2;q 11.2). Treatment options include an aggressive surgical approach and radiotherapy, whereas the role of chemotherapy remains to be defined. The case is described of monophasic synovial sarcoma located in the hypopharynx and a review is made of the literature concerning this rare neoplasm.

Riassunto

Il Sarcoma Sinoviale (SS) è una neoplasia mesenchimale maligna che solitamente colpisce gli arti inferiori dei giovani adulti. Le localizzazioni nei distretti testa collo sono invece rare. Istologicamente si riconoscono una variante bifasica ed una monofasica più rara e di difficile identificazione. L'immunoistochimica gioca un ruolo fondamentale nella diagnosi. Un ruolo importante assume anche la citogenetica essendo sia la forma monofasica che la bifasica caratterizzate da una traslocazione reciproca t(x;18)(p 11.2;q 11.2). Il trattamento prevede un approccio aggressivo chirurgico e radioterapico mentre il ruolo della chemioterapia non è ancora ben definito. Riferiamo su un caso clinico di sarcoma sinoviale monofasico a localizzazione ipofaringea revisionando la letteratura su questa rara neoplasia.

Introduction

Synovial sarcoma (SS) is a malignant mesenchymal type neoplasm accounting for approximately 10% of all soft tissue sarcomas. Mainly young adults and adolescents are affected with a male-female ratio of 1.2:1 [1].

The neoplasm often originates in proximity to the major joints and burses, particularly the lower limbs, while other localizations, such as the retroperitoneum [2], pleura, lung [3 4], thorax and mediastinum [5] are unusual sites.

SS of head and neck are very rare and account for only 3% of all sarcomas [6 19].

The first case of SS of the neck was reported by Jernstrom in 1954 [22] since when only about 85 cases have been reported [6 7].

Two different histological types have been identified and are generally referred to as the classic biphasic type, composed of epithelial cells and a spindle cell with a growth pattern which may be either glandular or solid, and a monophasic type in which a single cellular component is dominant; furthermore, other histological types of SS exist which are poorly differentiated and have a much lower incidence.

The monophasic form is more difficult to diagnose and, in this case, differential diagnosis is particularly difficult as far as concerns fibrosarcomas, spindle cell carcinoma, haemangiopericytomas and malignant schwannomas [1 8 22].

The case is reported of a monophasic SS of the hypopharynx and the clinical and pathological features of this rare neoplasm are described stressing the importance of immunohistochemistry and ultrastructural investigations in confirming this type of diagnosis.

Case report

This 22-year-old male patient came to our attention in May 1999 complaining of pharyngodinia and the sensation of a foreign body in the hypopharynx.

Direct laryngoscopy revealed a non-ulcerated yellow-

ish-brown neoformation, of soft consistency, located on the lateral wall of the left piriform sinus.
Computed axial tomography (CAT) (Fig. 1) revealed a neoformation of oval shape with a diameter of 3.3 cm and which showed a cranio-caudal extension of 4.5 cm with a dyshomogeneous "contrast enhancement" following contrast medium, with no evidence of calcifications. Only one lymph node was visible. This was enlarged in volume, at level III, on the left side.
A biopsy of the lesion was collected at microlaryngoscopy and the histological diagnosis was "Poorly differentiated monophasic synovial sarcoma (G2/G3)".
Brain, chest, upper and lower abdomen CAT, using contrast medium, as well as Total Body Bone Scintigraphy scan failed to reveal any metastases.
The patient was, therefore, submitted, following tracheotomy, to removal of the neoplasm, via lateral pharyngotomy, according to Trotter, with selective removal of lymph nodes II-III. On-the-spot diagnosis was negative as far as concerns the metastases in the lymph nodes examined then later confirmed following inclusion.
The post-operative course was uneventful and the tracheotomy was closed by the 10th day.
Histological examination (Fig. 2) revealed a dense cell proliferation, comprising elements of a fused appearance localised in bundles interrupted by irregular bundles of hyalinised collagen tissue. No areas of necrosis were present. The fused cells showed poorly defined cytoplasmic borders, with a central nucleus containing small areas of chromatin without a nucleolus. The borders were not damaged and a pseudocapsule with focal microinfiltrations was present.
Mitotic count was performed on sections stained with haematoxylin & eosin (H&E) and the mitotic index was expressed as the number of mitotic figures at 40X on 10 HPF (High Power Field). The DNA-Ploidy cytofluorimetric assessment showed a diploid neoplasm with a DNA index: 1.00, proliferative fraction (phase S): 4.9%, variation coefficient: 4.4.
Immunohistochemical evaluations, performed using the development method with streptovidine-biotin showed diffuse positivity for vimentine, whereas negative immunoreactions for desmin, cytokeratins and Cam 5.2, and a slightly positive immunoreaction for EMA and AE1.
It was not considered necessary, in this case, to carry out electron microscopy and cytogenetic studies since these assessments are not available in our Division and, moreover, are used primarily in uncertain cases.
A total of 24 lymph nodes were isolated and examined, none of which presented metastases.
TNM staging was: pT1b N0 M0, G3, stage IIB.
Treatment was completed with post-operative radiotherapy for a total of 60 Gy. At follow-up, in July 2001, a node was detected in the left lung in the lingular site. The patient was submitted to a "WEDGE RESECTION in VATS" (Atypical Video-assisted resection in Thoracoscopy).
The lesion was 1.3 cm in maximal diameter and histological diagnosis was secondary localisation of SS.
At the last follow-up, in January 2003, including a thoracic, abdominal and cerebral CAT, the patient is disease free.

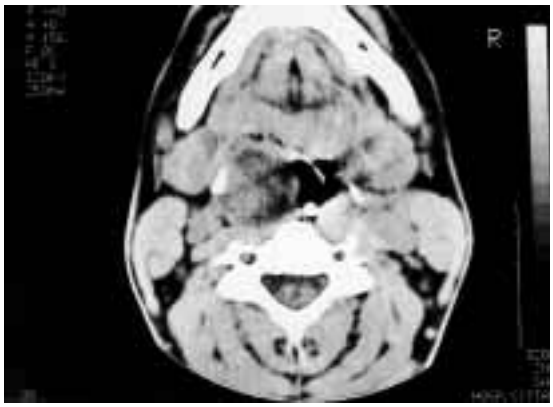

Fig. 1. CAT: axial scan.

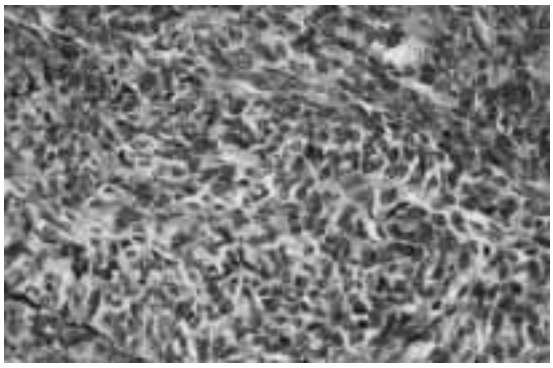

Fig. 2. Cell population rich in nucleolatus, roundish, monomorphic elements, showing a storiform pattern with focal hyalinisation (H&E. 250x).

Discussion and Conclusions

SS is generally considered a malignant tumour, with, biologically, an aggressive behaviour and with high probability of recurrence; in the head-neck region, this prevalence is very high and involves primarily elderly male subjects [6 8].

The neoplasm manifests as a indolent growing mass, the symptoms of which are correlated with the site of onset which, in decreasing order of frequency, comprises hypopharynx, neck, face, larynx, rhino- and retro-pharynx [9].

It usually occurs as a well-circumscribed mass covered by a thin fibrotic capsule making it easy to enucleate from the surrounding tissues. This is, however, a pseudocapsule beyond which the neoplasm usually tends to infiltrate [10].

Invasion of the tumoural pseudocapsule is usually accompanied by onset of distant metastases also in very distant districts. For this reason, simple enucleation is followed by recurrence in 90% of cases and, in radical exeresis of sarcomas of the limbs, free borders of at least 5 cm are necessary which, for anatomical limitations, are not feasible in the head and neck district [9]. This probably accounts for the high rate of local recurrences after limited exeresis.

CAT and nuclear magnetic resonance (NMR), whilst not specific, are useful in planning the surgical strategy. In 30-60% of SS cases, calcifications have been found at CAT examination and these, when present, are to be considered a positive prognostic factor [6 8 11].

The factors associated with a worse prognosis include: age >25 years, tumour size >5 cm and low grade of differentiation [9].

The probability of local recurrences has been reported to range between 30 and 40%, with 50% probability of distant metastases [10].

Onset of recurrences and metastases may manifest even after a prolonged period of time and, indeed, reports have appeared in the literature of recurrences even after 64 months [8]. Long-term follow-up, at short intervals [8], has been suggested which, in our case, included CAT evaluation of the brain and abdomen every 6 months besides routine clinical and endoscopic monitoring of the head and neck neoplasm. We carry out scintigraphic bone scans only in those cases with specific symptoms.

The most frequent site of metastases is the lung (49%), followed by skeleton (24%), liver (14%) and brain (11%) [9].

Metastatic spread occurs via blood whereas regional lymph node involvement is fairly limited, which, according to Cihak et al., is 12% [11] and prophylactic lymph node dissection is not advised in the presence of the N0 neck [6].

Survival at 5 years ranges between 23.5% and 45% [8 12] and between 11.2% and 30%, at 10 years [8 13].

From a histopathological viewpoint, SSs of the head and neck district differ from those of the limbs which would originate from a malignant transformation by cells of a mesenchymal nature [11].

SS, classically, show a biphasic cellular pattern comprising two cellular components: spindle cell and epithelial cells [1 8 9 11 14 22]: monophasic variants have also been described (epithelioid or fusiform) [11]. The pure monophasic epithelial pattern, as also the monophasic histotype with rounded cells, are somewhat rare and represent a real diagnostic problem particularly as far as concerns differential diagnosis [1]; the use of immunohistochemistry, in these cases, is a very important tool.

Positivity for cytokeratins in epithelial-like areas and the positivity for vimentine in mesenchymal-like areas with fused cells are crucial for diagnosis. Cases have been described, in the literature, in which both histotypes were positive for cytokeratins even if the number of positive fused cells was usually low [1 6].

Electron microscopy revealed the nature of this tumour which, despite the contradictory term of "synovial ", is derived, instead, from pluripotent mesenchymal cells; this would explain the possible onset in, not exactly, synovial sites [6 11 15].

In SS, a specific translocation exists between X and 18 chromosomes (X;18) (p11.2;q11.2).

The short arm (p) of chromosome X at 11.2 has translocated in the 11.2 region on the long arm (q) of chromosome 18. The finding of this translocation in the cytogenetic assessment is of considerable help in diagnosis, particularly in the less differentiated forms [1 11 19].

The American Joint Committee of Cancer classified all high grade sarcomas independently of the phenotype.

Cagle et al. found that SSs with a low mitotic index have a better prognosis, both in terms of survival and in disease-free period. The monophasic lesions, with a low glandular component and/or high mitotic index were associated with a worse prognosis [16].

Surgery remains the treatment of choice even if the probability of local recurrence is very high, 40-80% in adults and almost 90% in childhood. Use, therefore, of post-operative radiotherapy is, of course, advisable in all cases, with the exception of Grade I, and appears to improve local control; chemotherapy has been shown to have no impact on survival [6 9].

The role of chemotherapy in the treatment of head and neck SS appears to give rise to some controversy and does not seem to have a significant impact on disease-free survival [9 11], a positive effect would, on the other hand, appear to have been demonstrated for high grade sarcomas of the limbs [17]. Chemotherapy does not appear to have any impact on the control of later failure [18].

Patients with poorly differentiated SS (Grades II-III) show better survival when submitted to surgical treatment with post-operative radiotherapy and chemotherapy [9].

The few cases reported in the literature do not allow any definitive conclusions to be drawn concerning the most suitable treatment of soft tissue sarcomas located in the head and neck.

In our opinion, however, it appears worthwhile focusing on some observations:
– extended exeresis of the tumour with adequate volumes of normal surrounding tissue is not always possible in this district;
– local recurrence is very frequent with an unfavourable prognostic impact;
– distant metastases are very frequent and are usually synchronous with local recurrences;
– almost all those patients surviving long-term had a diagnosis of Grade I SS and with the size of the neoplasm <5 cm;
– radiotherapy and chemotherapy alone show little efficacy [12].

In conclusion, SS is a rare, highly malignant, neoplasm in the head and neck area which may give rise to diagnostic difficulties, particularly in the monophasic variant. Immunohistochemistry often allows diagnosis to be made for which, in dubious cases, cytogenetic studies and electron microscopy may provide useful tools. An aggressive surgical approach and post-operative radiotherapy in grades II and III are the options of choice in the management of these cases since the role of chemotherapy remains to be defined.
A scrupulous follow-up offers the possibility of early identification of metastases which, as in the case reported here, may often be successfully submitted to surgical treatment.

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Differentiating Monophasic Synovial Cell Sarcoma of the Kidney from Other Tumors of Kidney and Review of the Literature. Tanushri Mukherjee. * Oncopathologist, Command Hospital, Kolkata, India.Â Sarcomas of the kidney could be a clear cell sarcoma or Monophasic Synovial cell sarcoma (MSC) of the kidney is a tumor which is rare in kidney and is associated with poor prognosis. Differentiating this from other tumours is through histology and immunohistochemistry because of its characteristic spindly tumor cells with no epithelial component and immunopositivity for bcl2, CD99 and TEL1.Â (2009) Primary synovial sarcoma of the kidney: A case report and literature review. Korean J Pathol 43: 274-278. 8. Kim DH, Sohn JH, Lee MC, Lee G, Yoon GS, et al. Abstract Synovial sarcoma (SS) is uncommon high grade soft tissue sarcoma, accounting for less than 10% of all head and neck sarcomas. Also, about 10% of SS occur within the Head & Neck. In the pediatric population, SS is an extremely rare head & neck malignancy. Reference. ALOTAIBI, Naif Hudaik K, et al. Synovial sarcoma of the hypopharynx in a pediatric patient: Case report.Â Case report â€“ open access. International Journal of Surgery Case Reports 28 (2016) 1â€“3. Contents lists available at ScienceDirect. International Journal of Surgery Case Reports. journal homepage: www.casereports.com. Synovial sarcoma of the hypopharynx in a pediatric patient: Case report. N.H. Alotaibi. a,âˆ—. Synovial sarcoma in the parapharyngeal space: case report and review of the literature. Auris Nasus Larynx, Vol. 26, Issue. 1, p. 91.Â Postoperative Radiotherapy for Synovial Sarcoma of the Head and Neck during Pregnancy: Clinical and Technical Management and Fetal Dose Estimates. Tumori Journal, Vol. 93, Issue. 1, p. 45.