Case report

Myxofibrosarcoma of the thyroid gland

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A R T I C L E   I N F O

A B S T R A C T

Introduction: Myxofibrosarcoma of the thyroid is exceptional: a Medline search found a single case report. We report a new case which raised diagnostic and therapeutic problems.

Observation: We report the case of a 74-year-old woman who presented with swelling of the left thyroid lobe and ipsilateral cervical lymphadenopathy. Total thyroidectomy with cervical lymph-node dissection was performed. Histological analysis diagnosed myxofibrosarcoma. Evolution was marked by rapid local recurrence, and chemotherapy based on doxorubicin and ifosfamide was introduced.

Discussion/conclusion: Head and neck myxofibrosarcoma is rare. MRI is essential and should always precede treatment. Diagnosis is histological. There is elevated risk of local recurrence after resection, accompanied by worsening tumor grade, whence the need for accurate diagnosis, appropriate treatment and regular MRI follow-up.

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1. Introduction

Myxofibrosarcoma is one of the most common forms of sarcoma in elderly subjects. Location is generally in the limbs [1], although a few cases have been reported in the trunk (12%), retroperitoneal space and head and neck region (3%). The risk of local recurrence after resection is 50%, with tumor grade and prognosis worsening at each recurrence, with risk of metastasis [2]. We report an exceptional thyroid location which raised problems of diagnosis and treatment.

2. Observation

A 74-year-old woman presented with left cervical swelling of 2 months’ evolution. Clinical ENT examination found left lateral cervical swelling in the thyroid projection area, extending up into the submaxillary region. The mass was painful on palpation, soft, nodular, poorly contoured, 6 cm on the long axis, with ipsilateral submandibular lymphadenopathies. Otherwise, ENT and somatic examination found no abnormalities. Radiologic assessment comprising ultrasound and cervical CT (Fig. 1a and b) found a tumoral process in the left thyroid lobe with ipsilateral submandibular lymphadenopathies. Total thyroidectomy with tumor resection and ipsilateral functional lymph-node dissection was undertaken, and revealed a nodular, poorly contoured mass with gelatinous and fibrous areas. Immediate postoperative course was free of complications.

Histology (Fig. 2) found malignant tumoral proliferation comprising areas of cells with highly nucleolated hyperchromatic anisokaryotic nuclei and eosinophilic cytoplasm of varying abundance, with myxoid remodeling and numerous vascular sections. On immunohistochemistry (Fig. 3), the tumor cells were locally positive for anti-EMA (epithelial membrane antigen) antibodies. These morphological and immunohistochemical aspects indicated high-grade myxofibrosarcoma.

Extension assessment was negative. Control MRI at one month found a tumoral process infiltrating the parotid region with infratemporal and spinal extension suggestive of recurrence.

Six chemotherapy courses were instituted, comprising doxorubicin (75 mg/m²) and ifosfamide (7.5 mg/m²) every 3 weeks, supported by growth factors. Digestive and hematologic tolerance was good, but left ventricle ejection fraction was impaired (30%). Post-chemotherapy assessment could not be performed, as the patient was lost to follow-up.

3. Discussion

Soft-tissue sarcomas account for 1% of cancers in adults. Their rarity, ubiquity and morphologic variability often raise problems of diagnosis [1].

Myxofibrosarcoma is one of the most frequent sarcomas of the limbs in elderly subjects of both sexes [2], but is rare in the head and neck region, with only a few cases reported in the sphenoid sinus, maxillary sinus, mandible and hypophysyx [3].
MRI is essential and should systematically precede treatment [1]. In the present case, the radiological assessment alone was insufficient, falsely suggesting a large left thyroid nodule so that complementary examinations were not prescribed.

Diagnosis of soft-tissue sarcoma is essentially histopathological. Myxofibrosarcoma was first distinguished from malignant fibrous histiocytoma 29 years ago by Angervall et al. on the basis of histological and immunohistochemical criteria [4]. Histologically, there are often alternating myxoid and more cellular areas [4]. The proportion of myxoid areas required to diagnose myxofibrosarcoma is controversial: some authors set the bar at 50% of tumor volume [5], while for others a 10% rate of typical myxoid areas is enough [6]. The spindle-shaped or stellate cells have hyperchromatic nuclei. There is commonly mitosis. Myxoid areas show cells with vacuolated cytoplasm suggestive of lipoblasts but which are in fact mucin vacuoles, often poorly contoured and not deforming the nucleus [7]. In the present case, histology indicated myxofibrosarcoma (with more than 50% myxoid areas); immunohistochemistry found tumor cells were focally positive for anti-EMA antibodies.

Histologic grade is important for assessing metastatic potential and the need for adjuvant treatment. It is founded on cell differentiation, mitosis index and degree of tumoral necrosis [8] and should not be performed before precise histologic characterization of the sarcoma [1]. Myxofibrosarcomas are classified in 3 grades [3]: the present case was of high-grade myxofibrosarcoma.

Regarding evolution, one half of cases show local recurrence within 5 years, regardless of histologic grade or the depth of the primary, and recurrence may be found for up to 10 years. With repeated recurrence, tumor grade worsens; 13% of intermediate and high-grade tumors show metastasis [7,9].

These histologic and evolutive features call for precise diagnosis and regular MRI follow-up.

In the present case, total resection and lymph-node dissection were performed. Given the histologic diagnosis and bad prognosis in case of incomplete resection, regular MRI follow-up was undertaken to enable early screening of any recurrence, such as did indeed occur. Chemotherapy was initiated, with doxorubicin and ifosfamide. Unfortunately, the patient was lost to follow-up after 6 cycles, having very probably died.

4. Conclusion

Myxofibrosarcoma is a histological and clinical form of soft-tissue sarcoma worth highlighting due to its misleading aspect and particular prognosis. Ultrasound and CT are poorly contributive, often delaying diagnosis and complicating management. MRI is essential for assessment, and should always precede biopsy. Ideally, such cases should be discussed in regular multidisciplinary team meetings of all those concerned by this type of pathology.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References