wjpmr, 2023, 9(12), 05-07

SJIF Impact Factor: 5.922

WORLD JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.wjpmr.com

Case Study

ISSN 2455-3301

WJPMR

PULMONARY SYNOVIALOSARCOMA: A CASE REPORT AND REVIEW OF THE LITERATURE NATIONAL INSTITUTE OF ONCOLOGY RABAT

Ouail Benzerouale*[1], Fatima-Zahra Kahouadji[1], Sihame Lkhoyaali[1], Sarah Naciri[1], Saber Boutayeb[1], IbrahimEl Ghissassi[1], Hind M'rabti[1] and Hassan Errihani[1]

[1]Medical Oncology Department at the National Institute of Oncology in Rabat Morocco.

***Corresponding Author: Dr. Ouail Benzerouale**

Medical Oncology Department at the National Institute of Oncology in Rabat Morocco.

Article Received on 04/10/2023 **Article Revised on 25/10/2023** **Article Accepted on 15/11/2023**

ABSTRACT

Synovialosarcoma is a rare mesenchymal tumor with a poor prognosis. It accounts for 8% of soft tissue sarcomas. Its diagnosis is facilitated by immunohistochemical methods, but above all by molecular biology. It usually develops in the limbs, but its pulmonary localization remains exceptional. We report a clinical case of synovialosarcoma of the lung in a 54-year-old woman, discovered at a localized stage. This extremely rare tumor presents a particular immunohistochemical phenotype, which strongly contributes to the diagnosis. Cytogenetic studies confirmed the diagnosis, showing the presence of the characteristic specific translocation t (X; 18). This article highlights the diagnostic, therapeutic and prognostic features of this rare tumour, which is often overlooked by clinicians.

KEYWORDS: Synovialosarcoma, lobectomy, isofosfamide.

INTRODUCTION

Synovialosarcomas are malignant soft-tissue tumors located mainly in the limbs and nearby of large joints. They are high-grade, characterized by local invasion and a strong metastatic predisposition.[1]

The overall incidence of soft tissue sarcomas is estimated at 3 to 4 per 100,000 population, and 50 to 60% of these cancers develop in the limbs.[2]

They are more common in adolescents and young adults (15-40 years), with a sex ratio (3/1) favoring the male sex (1).

There are three anatomopathological forms: monophasic with spindle cells, biphasic with a double epithelial and spindle cell contingent, and finally the undifferentiated form.[3-4]

Primary thoracic sarcomas are rare, accounting for less than 1% of all primary thoracic tumours.[2]

Its multidisciplinary management requires the involvement of radiologists, pathologists, orthopedic surgeons, oncologists and radiation therapists.

Rare cases of synovialosarcoma with a pulmonary primary site have been described in the literature.

We report a new case and present the particularities of this tumour, which is rarely encountered in clinical practice

PATIENT AND OBSERVATION

Observation

Mrs. K.F, 54 years old, with no particular history of the disease, developed a chronic cough with purulent sputum complicated by hemoptysis and chest pain 7 months after diagnosis.

A thoracic CT scan revealed a process in the upper left lobe of the lung (6x6x7cm),

A scan-guided biopsy with additional immunohistochemistry was carried out, confirming a Grade II monophasic synovialosarcoma with positive anti-EMA marker and negative CD34, CD20, PS100 and anti-cytokeratin type AE1/AE3 markers.

The search for a fusion transcript specific to the t (X, 18) translocation was positive using the FISH technique.

The patient underwent neoadjuvant AI chemotherapy, followed by left upper lobectomy with lymph node dissection.

Anatomopathological examination of the surgical specimen showed a grade II spindle cell sarcoma (synovialosarcoma) classified pT3N0Mx, with healthy surgical margins and no vascular emboli or peri-nervous sheathing.

The patient then underwent three-dimensional adjuvant radiotherapy with a total dose of 66 Gy; 2 Gy/fraction in 33 fractions.

Six-month follow-up showed a good clinical-radiological response.

DISCUSSION

Synovialosarcoma is a highly aggressive soft-tissue malignancy of unknown pathogenesis, accounting for 7-8% of malignancies of mesenchymal origin.[5]

It occurs most frequently in adolescents and young adults, with an average age of 38.[5]

It occurs mainly in the limbs and nearby of large joints, primarily the knee.[5]

Thoracic involvement is varied, involving the heart, lung, mediastinum, esophagus, pleura and chest wall.[5]

Primary thoracic sarcomas are very rare compared with metastatic sarcomas.[5]

Clinically, patients with thoracic synovialosarcoma usually present with progressively worsening chest pain, cough or dyspnoea.

Synovialosarcoma of the chest wall presents as a soft lump, progressively increasing in size over several months or years.[5]

For our patient, no parietal mass was found.

Radiologically, synovialosarcoma is characterized by heterogeneity, sometimes containing calcifications in 25% of cases. Computed tomography can better assess the presence of micro-calcifications, as well as endothoracic and parietal extension.[2]

However, on magnetic resonance imaging, synovialosarcoma has a well-limited, capsule-like appearance, with partitions frequently present. The tumors are heterogeneous on T2, with liquid, solid or fibrous tonality, and sometimes the presence of intra-tumoral necrosis.[5]

Synovialosarcoma of the chest wall is rarely associated with liquid pleural effusion and, to our knowledge, only one case has been described previously.[5]

The usefulness of positron emission tomography (PET-CT) has been little studied, and bone scintigraphy is recommended in the event of a bone call point.[2]

For our patient, in the absence of extra-thoracic signs, the diagnosis of primary thoracic synovialosarcoma was evoked.

The diagnosis was finally confirmed by biopsy of the lung mass.

On pathological examination, the tumour was oval or rounded, sometimes multi-nodular, often well-demarcated and encapsulated, pale, whitish or greyish in color and soft in consistency.

Three subtypes of synovialosarcoma can be distinguished: the monophasic form (31%), which is a pure fibrosarcomatous form, the biphasic form (33%), which combines epithelial and spindle cells, and the undifferentiated form (36%), which contains small, oval or spindle-shaped cells characterized by sparse cytoplasm and dense nuclei.

On immunohistochemistry, synovialosarcomas express epithelial membrane antigen (EMA) and cytokeratins in 90% of cases, CD99 in 60% and S100 protein in 30%.[6]

Histologically, the majority of parietal synovialosarcomas reported in the literature present a biphasic aspect associating an epithelial contingent (expressing epithelial markers, cytokeratin and epithelial membrane antigen) and a mesenchymal contingent (expressing vimentin) with spindle cells circumscribing the epithelial structures.[7]

In most cases, there is a characteristic t (X; 18) translocation involving the SSX1 or SSX2 genes on the X chromosome (Xp11). Transcripts of the SYST-SSX fusion gene can be detected on pathological specimens with a sensitivity of 96% and a specificity of 100%.[8]

In our patient, immunohistochemistry revealed anti-EMA positivity and anti-cytokeratin negativity. The search for the t (X, 18) fusion transcript was positive using the FISH in situ hybridization technique.

The National Federation of Cancer Centers (FNCLCC) histopronostic score, which integrates 3 histological parameters: degree of tumor necrosis, degree of differentiation and percentage of mitoses.[2]

This score, applicable to the vast majority of soft-tissue sarcoma histologies, constitutes a powerful and easily reproducible prognostic factor, enabling sarcomas to be separated into low-grade (grade I), intermediate-grade (grade II) and high-grade (grade III) tumors.[2]

Therapeutically, sarcomas of the chest wall present a real problem in terms of therapeutic management, and in particular require wide surgical resection, the treatment of choice for synovialosarcoma to reduce the risk of locoregional and distant recurrence.[5]

This wide resection should be followed by restoration of chest wall stability and function.[5]

The advantage of adjuvant radiotherapy is that it enables better local control of the tumour. It is indicated when the tumour is 5 cm in diameter or more, or when the margins are insufficient.[5]

No study has evaluated the benefit of adjuvant chemotherapy in this situation.[2]

Treatment with Doxorubicin and/or Ifosfamide is the first-line therapy in inoperable and metastatic forms, with a response rate of around 50%.[2]

The average rate of locoregional or metastatic recurrence at two years is 50%.[2]

A tumour diameter of less than 5cm, a low mitotic index (Ki 67 < 10%), the absence of tumour necrosis and the absence of residual tumour after surgical resection are considered to be good prognostic factors.[2]

Survival at 5 years varies between 35% and 76% depending on the absence or presence of good prognostic factors respectively.[2]

In our case, the patient had a tumor size of 7 cm and a grade II tumor. Neoadjuvant chemotherapy accompanied by wide resection followed by adjuvant radiotherapy contributed to tumor regression and the favorable evolution of his disease.

CONCLUSION

Synovialosarcoma of the lung is a rare, highly aggressive malignancy.

Its diagnosis is difficult and requires immunohistochemical and cytogenetic analysis to distinguish it from other tumors. It can metastasize to the thorax, and much more rarely, originate in one of its compartments. L. Resection surgery, with or without tumor bed irradiation, is the standard treatment for operable tumors. In metastatic forms, ifosfamide-based chemotherapy improves prognosis, given the chemosensitivity of these tumors.

REFERENCES

1. Lukulunga LU, Moussa AK, Mahfoud M, El Bardouni A, Ismail F, Kharmaz M, Berrada MS, El Yaacoubi M. Résultats du traitement du synovialosarcome des members [Results of treatment of synovial sarcoma of the limbs]. Pan Afr Med J., Aug. 28, 2014; 18: 343. French. doi: 10.11604/pamj.2014.18.343.4293. PMID: 25574319; PMCID: PMC4282802.
2. Remy O, Messouna M, Akimana G, Kamdem M, Errihani H. Le synovialosarcome du poumon: à propos d'un cas et revue de la littérature [Pulmonary synovial sarcoma: case study and literature review]. Pan Afr Med J., Jun 29, 2020; 36: 137. French. doi: 10.11604/pamj.2020.36.137.23034. PMID: 32849992; PMCID: PMC7422737.
3. Eiber FC, Dry SM. Diagnosis and management of synovial sarcoma. J Surg Oncol, Mar 15, 2008; 97(4): 314- 20. PubMed| Google Scholar.
4. Frazier AA, Franks TJ, Pugatch RD, Galvin JR. From the archives of the AFIP: Pleuropulmonary synovial sarcoma. Radiographics, May-Jun, 2006; 26(3): 923-40. PubMed | Google Scholar
5. (L. Fekih). 0761-8425/$ — see front matter © 2011 SPLF. Publié par Elsevier Masson SAS. Tous droits réservés. doi:10.1016/j.rmr.2011.03.012.
6. Qassimi L, et al. Une tumeur rare de la paroi thoracique: le synovialosarcome. Rev Pneumol Clin., 2015. http://dx.doi.org/10.1016/j.pneumo.2014.12.004
7. Dos Santos NR, De Bruijn DR, Van Kessel AG. Molecular mechanisms underlying human synovial sarcoma development. Genes Chromosomes Cancer, 2001; 30: 1—14.
8. Ngahane BH. Évaluation des facteurs pronostiques du synovialosarcome thoracique. Rev Mal Respir, 2010; 27: 93—7. [éditorial].