heterogeneous intravescical mass, located at the posterior and left wall of the urinary bladder, that was confirmed by cystoscopy. The patient underwent transurethral resection (TUR) of the tumor that revealed a highly undifferentiated neoplasm comprised of highly atypical large cells with eosinophilic to vacuolated cytoplasm and hemophagocytosis. A wide panel of immunohistochemistry was set up and erroneously it oriented towards a sarcomatous origin for the tumor cells. On the basis of these results, a cystoprostatectomy was performed. Histologically, the neoplasm cells were obscured by a prominent inflammatory infiltrate consisting of small lymphocytes, plasma cells, benign histiocytes neutrophils and eosinophils. Occasionally haemophagocytosis was observed in association with vascular invasion. The neoplasm infiltrated the external muscular layer of bladder and consisted of epithelioid and pleomorphic cells with abundant eosinophilic cytoplasm often with some fine vacuoles. These malignant cells show large, round to oval nuclei, sometimes placed at the periphery with vesicular chromatin. In consideration of this unusual neoplasm histological pattern, it have been necessary a large immunohistochemistry determination. The immunohistochemical panel showed positive in tumoral cells including: CD68 (KP1 and PGM1), CD31, CD4, CD43, LCA, MPO and CD45. Negative stain was observed for GATA3, EMA, p63, ALK, CAM5.2 and S-100. The neoplastic cell showed a proliferative index Ki67 about 90%. Considering morphology and immunohistochemical staining profile, a diagnosis of HS was achieved. Shortly the patient developed pulmonary and liver metastasis and a recurrent mass in the pelvic wall occurred. His conditions got worse and died 2 months following the histologic diagnosis.

Results. HS is an extremely rare aggressive haematological neoplasm with very few numbers of reported series. These neoplasms are almost impossible to diagnose only by morphology because they share similar histologic feature with other neoplasm like epithelioid sarcoma, melanoma and carcinoma. For these similarities the use of immunohistochemistry markers is essential to provide a correct diagnosis of HS that shows the expression of specific histiocytic markers, as CD68 and lysozime. As it is reported in last WHO of 2017, the patients affected by this neoplasm has a wide age range, but most of the cases occurs in adult and some studies had found that there is a male predilection. The clinical presentation of this neoplasm is usually as localized mass, in rare case the neoplasm can present as disseminated diseases. In literature cases of brain, mediastinum, uterine cervix, choroid and liver localization have been reported. There is also a case report of HS in a patient with history of kidney transplant with multiple masses in native kidneys and liver. In WHO of 2017 is reported an association with metachronous or prior low-grade lymphoma (commonly follicular lymphoma), but also with chronic lymphocytic leukaemia/small lymphocytic lymphoma. The prognosis of patients affected by HS is very poor and it depends on stage and primary localization, the only treatment agreement seem to be surgical excision followed by chemotherapy and/or radiotherapy, but there are only few data.

Conclusions. To the best of our knowledge there is only one other report of bladder involvement by HS in a 80 years old man with a previous history of DLBCL. In our patient there is no previous history of lymphoma, transplantation or other neoplasm, so this is the first described case of primary HS in bladder without previous or metachronous lymphoma.

HS are very rare tumours and represented than 1% of all haematological malignancies, histological diagnosis is very difficult and cannot be diagnosed without auxilio of immunohistochemestry.

The use of histiocytic lineage such as CD68 (PGM1 and KP1) as diagnostic marker of HS must be considered for differential diagnosis in this very rare and difficult cases.

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ZINNER SYNDROME: A WOLFFIAN CONUNDRUM. A SYSTEMATIC REVIEW

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Background. Zinner's Syndrome consists in an aberrant development of the mesonephric duct and in the absence of the ureteric bud during embryogenesis, that leads to ipsilateral renal agenesis and atresia of the ejaculatory duct, which subsequently progresses to cystic dilation of seminal vesicles. Often It is considered to be the male counterpart of Mayer-Rokitansky-Kuster-Hauses syndrome seen in females.

The close embryologic relationship between the genital and urinary tracts could explains the developmental aberrations leading to this anomaly: an insult occurring before the 7th gestation week leads to maldevelopment of the distal part of the mesonephric duct producing atresia of both ejaculatory duct and the ureteric bud.

Materials and methods. Scientific literature is limited to case reports and small series; Pubmed research for Zinner Syndrome discosed a total of 90 items. The research has been restricted to English written full papers for the period 1995-2019. A total of 32 papers have been reviewed considering 48 patients including two cases of our observation.

Results. The syndrome is usually diagnosticated between the 3rd and 4th decades the main symptoms of presentation were urinary (n=18), pelvic/abdominal pain (n=12), infertility/ejaculatory impairment (n=9) while fever (n=2) and incidental finding (n=8) were rarer. In 8 cases other variable malformation were associated and in 2 other cases also other urologic malignancies were present. 45 cases out of 48 underwent to surgery at the time of the diagnosis or for exacerbation of the symptoms. The two cases of our observation were both in the 4th decades come to medical attentention for lower back pain and as occasional finding. The pathologic exami-

nation of the surgical specimens showed a dysplastic structures including a large cystic mass, morphologically and immunophenotipically of possible mesonefric derivation (GATA3+, PAX8+, CK7+, focally PSA+ and focally PSAP+) and with a prevalent tubule-cystic component surrounded by smooth muscle fibers (SMA+; h-Caldesmon+) arranged in helical bundles. One case also showed minimal differentiation in renal direction including scant tubulo-glomerular structures.
Conclusions. Zinner's syndrome is considered rare condition in general presenting the 3rd- 4th decades of life for the vague and aspecific urological symptoms; just a smaller population is diagnosed in pediatric age as occasional finding during screening imaging. The pathological examination of the surgical specimens appear crucial for correct diagnosis of the syndrome. The histological features are poorly covered in literature and may represent a surgical, pathological and embriological conundrum. A thorough histological examination with a limited panel of immunohistochemistry markers is required for a correct diagnosis considering the existence of other associated malformation and the risk of malignancy,

POST-RADIATION ANGIOSARCOMA OF THE BLADDER: A TRUE PATHOLOGICAL DIAGNOSTIC CHALLENGE. TWO CASE REPORTS AND LITERATURE REVIEW

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Aims. Primary angiosarcomas of the bladder are rare tumors of middle-aged and elderly men that typically present with locally advanced disease. The association of angiosarcoma with therapeutic radiation has been previously described. Microscopically it consists of atypical anastomosing vascular channels in most reported cases. It is not uncommon for this tumor to dedifferentiate and form sheets of primitive cells, spindle cells, epithelioid cells, or a mixture of these cell types. Awareness of the histopathologic spectrum of angiosarcoma is important for the correct diagnosis.
Material and methods. We herein report two new cases of this neoplasm, both occurred after a pelvic irradiation for prostatic adenocarcinoma, highlighting the clinical presentation, histological features, immunophenotypic profile, and treatment. At the best of our knowledge, one of the two cases is the first described for which the diagnosis of angiosarcoma was made with a concurrent metastatic disease in the bone-marrow biopsy.
Results. The sections from the transurethral resection chips of the bladder, in both patients, showed a densely fibrotic subepithelial connective tissue with extension of the sclerotic tissue within the muscolaris propria. The mucosa was extensively ulcerated with an evident granulation tissue immediately beneath, intermixed with a focal areas of proliferation of small and medium sized vessels sometimes anastomizing, lined by a plumped endothelial lining without frank atypia. The superficial urothelium, where present, showed focal reactive atypia. Immunohistochemical positivity for vascular markers, including CD31, CD34, ERG, and factor VIII lead us toward the right diagnosis.
Conclusion. Angiosarcomas can occasionally involve the urinary tract. Primary angiosarcomas of the bladder are rare tumors of middle-aged and elderly men that typically present with locally advanced disease. (1,2). Pathologists should have a high index of suspicion in elderly men presenting with hematuria and a urinary bladder mass (3,4). The association of angiosarcoma with therapeutic radiation has been previously described (5). The absence of a history of radiotherapy does not exclude angiosarcoma. The broad pathologic spectrum of primary bladder angiosarcomas could make a right diagnosis extremely difficult, particularly on limited biopsy material (6). The pathology is that of typical angiosarcoma in only half of the cases. Awareness of the histopathologic spectrum of angiosarcoma is important for the correct diagnosis, especially when dealing with small biopsy specimens. The most useful morphological characteristics to raise one's suspicion of an angiosarcoma are the presence of blood-filled spaces of variable size and shape lined by atypical cells. This "classic pattern" with typical cytology was the most common pattern reported in primary bladder angiosarcomas (6). However, this finding may be focal and overlooked. Moreover almost half of the cases had a solid growth component, and roughly one third had either spindled or epithelioid cytology. Hence, immunohistochemical stains are often crucial to confirming the diagnosis of angiosarcoma. In atypical cases, testing with immunohistochemistry is essential. Immunohistochemical positivity for vascular markers, including CD31, CD34, ERG, and factor VIII should lead toward the right diagnosis(6,7). Most cases are negative for cytokeratin but focal positive cytokeratin stain is commonly seen and should not be interpreted as sufficient evidence of epithelial origin (6). Evidence of c-Myc amplification, as seen in the current case report, may be supportive, as it is found in over half of the post-radiation angiosarcoma (7).

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XP11 TRANSLOCATION RENAL CELL CARCINOMA DIAGNOSED IN BONE METASTASIS IN ADULT PATIENT: DESCRIPTION OF TWO CASES

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