CASE REPORT

SYNOVIAL SARCOMA – INGUINAL REGION A RARE FINDING

Muhammad Salman, Ishtiaq Ahmed, Shahid Rasul

ABSTRACT

A 25 years old male presented with suspicion of lymphadenitis over right inguinal region and exploration was inconclusive. Later on patient presented with recurrence and extensive ulcer at the same place which was diagnosed synovial sarcoma on histopathology. Case was managed by Neo-adjuvant chemotherapy followed by surgery.

KEY WORDS: synovial sarcoma, inguinal region, surgery, chemo therapy, radiotherapy

INTRODUCTION

A synovial sarcoma is a rare form of soft tissue cancer which was originally coined early in the 20th century as some thought that the microscopic similarity of some tumors to synovium and its propensity to arise adjacent to joints indicated a synovial origin[1]. Despite its name, synovial sarcomas do not appear to arise from synovial membranes but from unknown multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures[2,3] and lack synovial differentiation[4,5]. Synovial sarcoma is the fourth most commonly occurring sarcoma, accounting for 7-8% of all sarcomas[6]. It most frequently occurs in young adults, with up to 30% being manifested during the first two decades of life, and a median age of 13 years at presentation[7,8,9]. With an annual incidence of around 2-3/100,000, they account for less than 1% of all malignant tumors and 2% of all cancer-related deaths, although in children soft tissue sarcomas represent about 8% of all malignancies[1,7,8]. The peak of incidence is before the 30th birthday and males are affected more often than females (ratio around 1.2:1) [7]. We are presenting a case of synovial sarcoma over inguinal region initially misdiagnosed as lymphadenitis.

CASE HISTORY

As per previous documents this 25 years old male labour presented with one month history of painless swelling over right inguinal region. There was no history of fever, trauma or any other complaint. Clinical examination showed a 1 cm diffuse, rounded, firm to soft swelling over right inguinal region. Overlying skin was comparatively erythematous as compared to surrounding skin. There was no fluctuation, transillumination or cough impulse. Provisional diagnosis of inguinal lymph adenitis/abscess was made by a local doctor who planned incision and drainage/excision biopsy. As per record incision was closed without any excision or biopsy because exploration by operating medical officer was inconclusive. The patient reported our OPD 13 days after initial surgery with wound infection for which drainage was performed and the wound kept open for daily dressings which was followed by complete resolution. About two month later patient again noticed a swelling at the same region which was rapidly expanded into a fungating lesion. Incision biopsy was done which was suggestive of synovial sarcoma and immunohistochemistry showed CD95 - positive, EMA - focal positive, CKAE/AE3 - negative and LCA-negative. Patient was advised neo-adjuvant chemotherapy followed by surgical excision and repair of defect through myocutaneous flap.

Fig. 1: Extensive fungating lesion over right inguinal region.

Correspondence to:
Prof. Dr. Ishtiaq Ahmed
Consultant Surgeon
Al-Nafees Medical College & Hospital
Isra University, Islamabad Campus, Pakistan
E-mail: surgish2000@yahoo.com

DISCUSSION

Primary synovial sarcomas are most common in the soft tissue near the large joints of the arm and leg but have been documented in most human tissues and organs including the eye [10], heart[1] duodenum [7] liver[8],pharynx [11], kidney [12], lungs, pleura [13] prostate, brain [12], vulva [5,14] etc. Pathogenesis of synovial sarcoma is still unknown and there are no well-established risk factors. Therefore no screening program has been evaluated or could be recommended[6]. Synovial sarcoma is clinically, morphologically and genetically a distinct sarcoma characterized by the specific chromosomal translocation t(X; 18) (p11; q11). In the last WHO Classification of soft tissue and bone tumors, synovial sarcoma is classified among malignant tumors of uncertain differentiation lacking a precise normal tissue counterpart [3,7,15].

Synovial sarcoma usually presents with an otherwise asymptomatic swelling or mass. Symptoms may be related to these locations, although the painless mass remains the most frequent presentation. Difficulty in swallowing and breathing or alteration of voice might be associated with synovial sarcoma of the head and neck region. Pain may be related to the involvement of nerves. Because the tumor grows slowly, symptoms may be present for a long time before the diagnosis is made, which may delay the diagnosis. Although general symptoms related to malignancies can be reported such as fatigue [16,17] etc.

In order to determine local and distant extent of disease, a complete set of radiological investigations are necessary at diagnosis. Imaging studies are critical in determining tumor size and local extent. Ultrasonogram is often the first instrumental assessment to be used. Computer Tomography scan or MRI of the primary site are mandatory for local extension assessment before any treatment. MRI of an extremity is usually considered superior in defining soft tissue extension [17,18].

After the accurate description of the local extent of the tumor, pathological assessment is necessary to define the histological diagnosis. The initial biopsy has the aim to define the diagnosis but also should provide enough material for immunochemistry and cytogenetic biological studies [19]. Open biopsy (Incisional biopsy) or core needle biopsy (tru-cut, guided by ultrasound or CT scan) is preferred to fine needle aspirates that could establish the presence of malignancy, but rarely identify the subtype or provide the tissue required for additional studies. In any case, the initial biopsy should be carefully planned by an experienced surgeon taking into account the possible subsequent definitive surgery, which must include the scar and the biopsy tract [19,20].

After the radiological evaluation of tumor extent and biopsy, the diagnostic work-up is completed by staging investigations, aimed to detect regional and distant metastases. Chest CT scan, Technetium bone scan and abdominal ultrasound are required to identify lung, bone, and abdominal metastases. Attention to regional lymph nodes of the primary site is also important. Positron emission tomography is not yet considered a standard staging investigation for synovial sarcoma [1] Synovial sarcoma is a soft tissue sarcoma and is divided into three subtypes: biphasic, monophasic and poorly differentiated [6,7]. The biphasic type contains epithelial and spindle cell elements, which occur in varying proportions, whilst the monophasic type contains only spindle cells [1,14]. The poorly differentiated type shares features of both the monophasic or biphasic types and has varying proportions of poorly differentiated areas characterised by high cellularity, pleomorphism, numerous mitoses and often necrosis. All morphological subtypes are characterised by a specific t(X; 18) (p11.2; q11.2) chromosomal translocation [3,5].

A risk-adapted treatment strategy is based on pre-treatment staging and post-surgical clinical status. Pediatric oncologists usually stage synovial sarcoma according to the clinical TNM classification of malignant tumor (based on local invasiveness, T1 and T2, and tumor size, A or B, i.e. less or more than 5 cm; absence or presence of nodal and distant involvement: N0 and N1, M0 and M1, respectively [21]. The Intergroup Rhabdomyosarcoma Study (IRS) post-surgical grouping system is based on the degree of surgical resection [22]

- Group I - completely-excised tumours with negative microscopic margins
- Group II - grossly-resected tumours with microscopic residual disease and/or regional lymph nodal spread
- Group III - gross residual disease after incomplete resection or biopsy
- Group IV - metastases at onset

Adult oncologists often stage using the American Joint Committee on Cancer AJCC staging system which incorporates the histological grade (which is presumably high in all synovial sarcoma patients) in addition to size and depth (most SS are deep-seated). [17,21]

Aim of surgery is to remove the tumor and a safety margin of healthy tissue. This is the mainstay of synovial sarcoma treatment and is curative in approximately 20-70% of patients depending on the particular study being quoted [6]. Often synovial sarcoma has a pseudocapsule that allows the tumour

to shell out fairly easily, giving a false sense of security that complete removal has occurred [5]. The presence of positive microscopic margins gives a greater likelihood of local recurrence and is associated with an increased risk of metastatic spread and decreased length of disease-free survival. Therefore, the surgeon should aim at obtaining negative margins, although there is no clear evidence on the extent of negative margin required. Primary re-excision should be carefully considered in all patients with a gross residual tumour, as re-excision decreases the risk of local recurrence [5,23].

After diagnostic biopsy, chemotherapy and/or radiotherapy should be given to shrink the tumor and make it more amenable to subsequent surgery. [23].

The June 2004 Milan Consensus Conference on adult soft tissue sarcomas suggested a definition to which adequate margins are "those > 1 cm of healthy tissue around the tumor, in all directions, when the tissue is a muscle, and > 1 mm of healthy tissue around the tumor when the tissue is periostium, vessel sheath, epineurium or muscular fascia." Inadequate surgical margins adversely affect local outcome and consequently also overall survival though some studies on adult soft tissue sarcomas failed to find a strong correlation between quality of surgery and final outcome [23,24].

Conventional chemotherapy (for example doxorubicin hydrochloride and ifosfamide) is used to reduce the number of remaining microscopic cancer cells. The benefit of chemotherapy in synovial sarcoma to overall survival remains unclear, although a recent study has shown that survival of patients with advanced, poorly differentiated disease marginally improves with doxorubicin/ifosfamide treatment [25].

Radiotherapy aims to reduce the chance of local recurrence. The benefit of radiotherapy in this disease is less clear than for chemotherapy. Radiotherapy appears to have a role in the treatment of patients newly diagnosed with synovial sarcoma, especially children with minimal primary tumour or following surgery. Radiotherapy should improve local control of the disease but does not affect overall survival. In adult patients with soft tissue sarcoma, radiotherapy is usually recommended after incomplete resection, but often also after wide excision, especially in case of large tumour [26]. Certainly, the indication for radiotherapy has to be stricter in children and young adolescents with synovial sarcoma than in adults given the higher risk of severe late effects of radiotherapy. The role of adjuvant chemotherapy in the management of synovial sarcoma remains controversial [5].

The prognosis of synovial sarcoma patients is related to the feasibility of surgical resection, tumor size and local invasiveness. Patients with small tumors that can be completely removed at diagnosis have an excellent prognosis. For tumors larger than 5 cm, the risk of developing distant metastases is higher. Pediatric series[16-18,27] reported survival higher than 80% for IRS Group I-II patients but around 60-70% for cases > 5 cm. For patients with unresectable disease at diagnosis (IRS group III), survival has been reported between 50 and 70% (but it clearly is less for cases localized in head-neck region, lung and mediastinum, abdomen). For patients with distant metastases, overall results are poor [7,28].

CONCLUSION

Synovial sarcoma is not uncommon entity and can be found anywhere in body. Any lump in body should be explored by experienced surgeon and biopsy should be sent to minimize the chances of misdiagnosis.

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