



Granular cell tumor in the inguinal canal: A case report and literature review

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Abstract Granular cell tumors (GCT) are rare soft tissue neoplasms that may be asymptomatic or may be presented as a slow growing nodule. The diagnosis is established on the basis of cytological and histopathological characteristics of the disease. We present a case of granular cell tumor in the right inguinal region misdiagnosed as inguinal hernia.

Case report:

A 49-year-old female presented with a tender firm swelling in the right inguinal, 3x3 cm, firm in consistency that was present for three years. The swelling was reducible and the cough impulse was positive. Laboratory investigations were normal. Blood analysis showed: WBC: 7000/u/l, Hb 11.5 g/dl, PLT 245 k/l, ESR 12 mm 1st hour uca 14 mg/dl, Cre .9 mg/dl, FBS 89 mg/dl, and urinalysis was normal. A clinical diagnosis of inguinal hernia and hydrocele of canal of Nuck was performed. Ultrasound examination suggested an inguinal hernia. At surgery, a cystic mass, suspected to be hydrocele of Knucke, was resected and sent for histopathology (Fig 1-4). Histopathology revealed sections with thick fibrous cystic walls, lined by one cell layer of simple mesothelial epithelium with foci of squamous metaplasia without evidence of atypia. The underlying stroma showed dense fibrohistiocytic reaction, along with many multinucleated foreign body giant cells. Unremarkable fragment of striated muscle was observed. There was no evidence of malignancy. The diagnosis was of a hernia sac associated with nodular histiocytic mesothelial hyperplasia.

Introduction:

Granular Cell Tumor (GCT) is a rare benign tumor arising predominantly in skin, subcutaneous tissues and tongue.¹ They may occur at any age, but commonly affects third to fourth decades of life. Generally, these are isolated lesions, but can be multiple in about 15% of cases.² These tumors usually behave in a benign fashion, but malignant variant can also occur de novo or from transformation of a benign one. Earlier GCT was proposed to be derived from immature skeletal muscle cells (hence, the term granular cell myoblastoma was given). However, later ultrastructure and immunohistochemistry studies have proven it to be of schwannian differentiation.¹⁻⁴

To date, the cause of Langerhan Cell Histiocytosis (LCH) has not been determined. Multiple potential causes have been explored, including viruses, molds, infections, genetics, geographic location, racial clustering, seasonal changes, and environmental exposure. Results of these studies are still insufficient to provide a definitive answer, although in recent years there has been increasing evidence that mistakes that occur in certain genes of blood cells, also called gene "mutations", are an important cause of LCH. These mutations are not "genetic" in the sense of being inherited and passed from parent to child (thesae are called "germline" mutations), rather they occur only in certain cells in the affected individual (and are called "somatic" mutations). In particular, there is a mutation in a gene called BRAF in approximately one-half of cases of LCH. Mutations in other genes in the same family as BRAF have been found as well.⁵⁻⁶

Langerhans cell histiocytosis (LCH) can involve nearly any part of the body, though some sites are more common than others. A patient may have very limited involvement in one body system or widespread involvement in several different sites

and systems. It is also possible to have LCH in a particular location without symptoms.

The following symptoms may indicate disease involvement, but are not diagnostic of LCH. This disease varies greatly from patient to patient, and only some of these symptoms may be present:⁵⁻⁷

- Skin symptoms (scalp, face, groin, trunk, armpits, arms, legs, ear canals, vulva, anal area, fingernails) Rash, ulceration, redness, pain, oozing, hair loss. Lesions may appear as small, solid, reddish elevations on the skin surface; knots under the skin; purplish-red spots; bleeding under the skin; rashes that are scaly and greasy; ulcerations; and small abscesses. Skin lesions may signal that other parts of the body are affected.

- Bones symptoms (skull, bones around the eyes, back bones, ribs, pelvis, feet, toes, hands, and fingers). Pain, swelling, lumps, headaches, spontaneous fracture, ulceration at the site, limp, collapse of disc in the back, and inability to walk.

- Bone Marrow Low blood counts (red blood cells, white blood cells, platelets/clotting cells).

- Lungs symptoms include chest pain, shortness of breath, collapsed lung, dry cough, weight loss, fatigue, loss of appetite, night sweats, and coughing up blood.

- Teeth/Gum symptoms loosening or loss of teeth, swollen or bleeding gums, ulcerations, pain, and swelling of face.

- Ear symptoms Chronic ear infections, drainage/discharge, balance problems, bleeding, and decreased hearing. Ear involvement can be a result of skull lesions expanding to the ear and may cause balance problems. Ear canals may be affected as part of the skin system, causing bleeding/drainage.

- Endocrine System (pituitary gland/diabetes insipidus, hypothalamus, and thyroid gland) Severe thirst, excessive urination, fatigue, sweats, temperature swings, weight gain, weight loss,

nipple tenderness/discharge, menstrual problems in women, and erectile dysfunction in men.

- Female Genital/Reproductive Tract (vulva, vagina, cervix, ovaries) Inflammation, rash, and/or ulceration of the vulva, vagina, and/or cervix, but it may also affect the ovaries, causing dysfunction or failure of these organs.
- Liver/ Spleen/ Lymph Nodes Swelling/tenderness of abdomen; enlargement of lymph nodes, spleen and/or liver; fatigue.
- Gastrointestinal System (stomach or intestines) Diarrhea, nausea, vomiting, and/or weight loss.
- Central Nervous System (spinal cord, brain lesions, pituitary gland) Staggering when walking, seizures, weakness of the arms, legs, or weakness of one side of the body. Since the pituitary gland

could be affected, lesions of this system may also cause some of the same endocrine problems mentioned above.

- Additional Non-specific Symptoms: Fever, weakness, fatigue, chronic pain, weight loss, and night sweats.

Pulmonary LCH (PLCH) can occur as part of multi-system disease, or it can occur alone. Nearly 20% of adults with PLCH do not have present symptoms. An estimated 10%-15% of patients present with lung collapse as the first symptom, while others may show abnormalities in lung function tests. Dry cough and shortness of breath are the most common complaints, and weight loss, fever, sweating, and loss of appetite occur in approximately 33% of patients.⁵⁻⁷



Fig. 1. Inguinal cystic mass – intact a



Fig. 2. Inguinal cystic mass – intact b



Fig. 3. Inguinal cystic mass exposed a



Fig. 4. Inguinal cystic mass exposed b

Many adults experience severe and sometimes overwhelming pain associated with this disease. While pain can be caused by bone lesions or bone defects that do not heal completely with therapy, it has also been observed that some patients suffer pain even when there is no active disease seen on x-ray. The cause of this pain is not understood; however, it is being currently discussed in research. Pain is considered to be a complication of LCH that should be fully evaluated by a physician and treated with appropriate medications.⁵⁻⁷

Discussion:

A wide range of pathologic conditions affect the groin, and radiologists need to be aware of the variety of these entities. Although many of these disease processes may commonly appear similar on sonography and may require a biopsy or fluid aspiration to confirm the diagnosis, sonography, nevertheless, can be helpful in the evaluation of some of these diseases by enabling the differentiation between cystic and solid tumors. When the diagnosis is somewhat problematic, CT can be used for further evaluation. Computed tomography is more specific than sonography for the diagnosis of lipomas, hematomas, and abscesses.⁶

Misdiagnosis, in our case, was due to several reasons, foremost being the site of occurrence, clinical and radiological explanation of the inguinal mass.

The diagnosis of Langerhans cell histiocytosis (LCH) was made following a biopsy and microscopic examination of the affected tissue. Histopathology revealed sections with thick fibrous cystic walls, lined by one cell layer of simple mesothelial epithelium, with foci of squamous metaplasia, without evidence of atypia. The underlying stroma showed dense fibrohistiocytic reaction, along with many multinucleated foreign body giant cells. Unremarkable fragment of striated muscle was observed. There was no evidence of malignancy. The diagnosis was of a hernia sac associated with nodular histiocytic mesothelial hyperplasia.⁷⁻⁹

If the biopsy of such mass is positive for LCH, some other tests are to determine the extent of disease and draw out the line of management. These may include skeletal x-ray survey with skull x-ray series, bone scan, blood tests (complete blood count, sedimentation rate, liver function tests, electrolytes), and urine tests. In case symptoms suggest diabetes insipidus, a water-deprivation test should be done. When the bones near the ears or eyes are involved, a head CT scan

is performed. An MRI of the brain may be necessary if the lesions are near the eyes or ears or there is suspicion of central nervous system (CNS) involvement. Chest CT and lung function studies are performed when indicated. Other testing may be done, depending on the symptoms. Other biopsies may be performed if test results or abnormal findings during the physical exam cause the physician to suspect involvement of another area.⁷⁻⁹

When lung involvement is suspected, a chest x-ray is usually performed and is abnormal in most patients. The most sensitive diagnostic test is a high-resolution CT scan (HRCT), another type of x-ray, which can reveal lung cysts that are characteristic of LCH. It is possible to make a diagnosis with a bronchoalveolar lavage (BAL). The BAL is a medical procedure in which a tube is passed through the mouth or nose into the lungs, and fluid is squirted into part of the lung and then collected for examination. If the BAL and HRCT do not provide a diagnosis, then surgical lung biopsy may be necessary. In patients with diagnosed LCH in other areas of the body such as skin or bone, the diagnosis of lung involvement can be made if the HRCT shows findings consistent with pulmonary Langerhans cell histiocytosis (PLCH).⁹

Treatment is based upon the organ(s) involved, extension of disease, and in some cases, age of the patient. In some cases, no treatment is necessary. Others may respond to surgical removal, as was for our patient. Other patients may require steroids, or anti-inflammatory drugs (NSAIDs). Low-dose radiation is helpful in some cases, but should be carefully used in children. There are patients who require chemotherapy, such as vinblastine, vincristine, etoposide (VP-16), methotrexate, cytosine-arabioside (Ara-C), and/or 6-MP. In patients with severe disease that does not respond to initial treatment, stronger chemotherapy combinations may be used. Ultraviolet light (PUVA) may be helpful in limited skin disease. In very rare instances, a transplant of the liver, lung, or bone marrow may be necessary.⁷

Even if it is determined that no treatment is necessary, apart from resection of the lump as was for our patient, at the time of diagnosis, the

physician should monitor the disease regularly to be sure that there is not further progression of the existing areas and that new infected areas can be detected and treated early. The ultimate goal of an overall treatment plan, of course, is to use as little treatment as possible to keep the disease under control to preserve quality of life and prevent the histiocytosis from damaging vital organs.⁷⁻⁹

Conclusion:

The possibility of granular cell tumor, although rare, should not be overlooked in the course of workup of patients with inguinal lump.

References:

- [1]- Pathania K, Bhargava S. Granular cell tumour of breast: A mimic of carcinoma. *MJAFI*. 2010;66:292-4.
- [2]- Lack EE, Worsham GF, Callihan MD, Crawford BE, Klappenbach S, Rowden G, et al. Granular cell tumor: a clinicopathologic study of 110 patients. *J Surg Oncol*. 1980;13:301-16.
- [3]- Ordonez NG. Granular cell tumor: a review and update. *Adv Anatomic Pathol*. 1999; 6:186-203.
- [4]- Rao S, Rajendiran S, Venkata PM, Prathiba D. Granular cell tumour in inguinal region: A case of mistaken identity on cytology. *J Cytol*. 2012; 29:60-62.
- [5]- Colin T, Blackburn AV, Milner RH, Gerrand C, Ragbir M. Sarcomas in the groin and inguinal canal – often missed and difficult to manage. *Ann R Coll Surg Engl* 2010; 92:
- [6]- Mo YD, Cheol KH, Wook J, Young KG, Hyuni C. Sonographic findings of groin masses. *Wiley Online Library*. [Wiley Online Library. 2007.](http://www.wiley.com/journal/10.1002)
- [7]- Histiocytosis Association. Langerhan Cell Histiocytosis in adults. <https://www.histio.org/page.aspx>. 2017.
- [8]- Liu K, Madden JF, Olatidove BA, Dodd LG. Features of benign granular cell tumor on fine needle aspiration. *Acta Cytol*. 1999; 43: 552-7.
- [9]- Vikas SK, Vyankatesh TA. Fine needle aspiration cytology of a subcutaneous granular cell tumor. *Bangladesh J Pathol*. 2010;25:23-24.