Primary ovarian non-Hodgkin's lymphoma in a 1-year-old child: A rare case report

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ABSTRACT

Primary ovarian non-Hodgkin's lymphomas (NHLs) are extremely rare. The definitive diagnosis can be done only by correlation of the clinical, radiological and pathological findings. Secondary ovarian lymphomas as a part of systemic disease are well known. We report a case of primary NHL of the ovary in a 1-year-old female child with unilateral involvement. Ultrasonography revealed a large pelvic mass, without associated regional or distant lymphadenopathy. The diagnosis was established by histological study and subsequent immunohistochemistry of the excised tumor mass. The case is rare due to its primary nature and presentation in this age group. Prognosis of primary ovarian lymphoma is the same as that of other primary nodal lymphomas. We want to highlight the importance of considering this as a differential diagnosis of lower abdominal mass in appropriate clinicoradiological scenario.

Key words: *Child, Non-Hodgkin's Lymphoma, Ovary, Primary*

Non-Hodgkin's lymphoma (NHL) can involve any part of the female genital tract, but the ovary is the most common site to be involved. The ovarian involvement in NHL is usually secondary, as a part of systemic disease [1,2]. The primary ovarian NHL (PONHL) is very rare accounting for 0.5% of all NHL and 1.5% of all ovarian neoplasms [3]. The patients of primary ovarian lymphoma usually present with pelvic complaints, and the size of the tumor is usually larger than the secondary lymphomas at the time of presentation [4]. The most common histological types of PONHL are Burkitt's lymphoma and diffuse large B-cell lymphoma [5]. We report a case of primary ovarian lymphoma in a 1-year-old female child which was diagnosed by histopathological examination and immunohistochemical study.

CASE REPORT

A 1-year-old female child presented with acute abdominal pain. Her personal and family history was unremarkable. General examination revealed mild pallor and increased temperature. There was no lymphadenopathy. On local examination, the abdomen was distended, tensed, umbilicus was everted. On palpation, there was an ill-defined mass in the right iliac fossa.

Emergency laparotomy was done, and the mass was resected. Pre-operative assessment, including complete blood count and peripheral blood smear, revealed no significant abnormality. The ultrasound abdomen showed a mass in the lower abdomen, probably arising from the lower gastrointestinal tract.

On gross examination, the specimen was a soft-tissue mass measuring (8×6×4) ccm with an attached fallopian tube at one side (Fig. 1). The surface was nodular and whitish in color. The cut section was predominantly solid homogenous whitish with small areas of hemorrhage and necrosis (Fig. 1).

Microscopic examination showed that the tumor was composed of small monomorphic round to oval cells, in solid sheets and cords, separated by fibrovascular septa (Fig. 2). The individual cells had scanty cytoplasm, clumped nuclear chromatin pattern and inconspicuous nucleoli (Fig. 2). A diagnosis of small round cell tumor was given with the advice of immunohistochemistry for further categorization of the lesion.

Subsequent immunohistochemistry revealed diffuse and strong positivity for leukocyte common antigen (CD 45) with a 4+ score (Fig. 3a) and negative for calretinin (Fig. 3b). It also shows diffuse positivity for B-cell marker, CD-20 (Fig. 4a), and negative for pan T-cell marker CD-3 (Fig. 4b). Post-operative bone marrow aspiration showed no abnormal cell. The patient was treated with five cycles of chemotherapy. Follow-up for 1 year showed no evidence of recurrence or metastasis.

DISCUSSION

Primary ovarian lymphoma is extremely rare. Only 0.5% of all NHL arise primarily from the ovary, and this accounts for about 1.5% of all ovarian neoplasms [3]. Other small round cell tumors such as rhabdomyosarcoma, teratoma, neurogenic tumor, granulosa cell tumor, dysgerminoma, undifferentiated carcinoma,

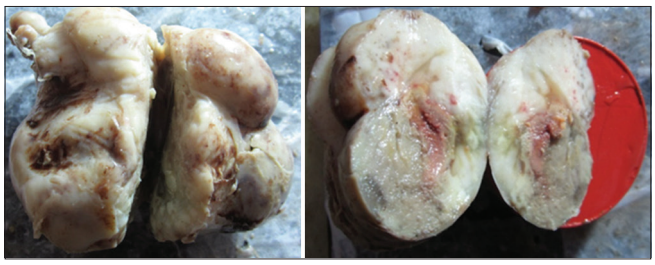

Figure 1: Gross examination and cut section of the specimen with attached fallopian tube at one side

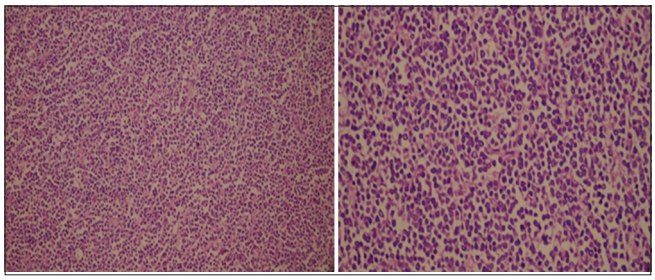

Figure 2: Low power and high power view of the tumor section (H&E)

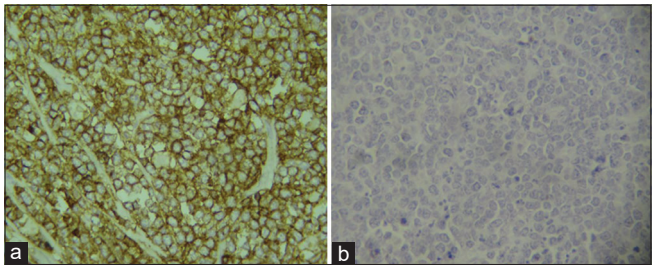

Figure 3: Immunohistochemistry (a) CD 45 positive; (b) calretinin negative

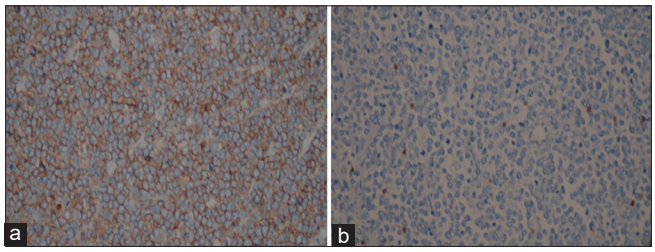

Figure 4: Immunohistochemistry (a) CD 20 diffusely positive; (b) CD 3 negative

and metastatic carcinoma are the differential diagnosis of PONHL. An exact diagnosis can only be done by histological examination of the tumor [6-8].

The possibility of the occurrence of primary lymphoma in the ovary was debated in the previous studies [9] because pre-existing benign lymphoid cells should be present in the ovary for the development of a primary lymphoid malignancy [1]. However, the presence of scattered B and T lymphocytes within the ovary was reported by Skodras *et al*. and Suzuki *et al*. in their studies [9,10]. These findings support the fact that primary non-Hodgkin lymphomas can arise even from the ovarian tissue [11].

In 1988, Fox *et al*. proposed the criteria for the diagnosis of PONHL [12]. The criteria were as follows: (a) The lymphoma should be confined to the ovary or the adjacent lymph nodes or structures at diagnosis, without evidence of lymphoma elsewhere; (b) the peripheral blood and bone marrow should not contain any abnormal cells; and (c) remote involvement should occur at least several months after ovarian involvement. In our case, the tumor mass was of ovarian origin and the hematological, as well as radiological reports of the patient, did not reveal any evidence of lymphoid malignancy at the time of diagnosis and even during follow-up period.

Primary ovarian lymphoma can occur at any age with a mean age of 35 years (range – 6–74 years) [1,13]. After extensive literature search over PubMed, very few cases presenting with PONHL were found, but interestingly, the age group is widely variable. Senol *et al*., in 2014, reported five cases of PONHL in older age groups (50–65 years) [14]. Bhartiya *et al*., in 2016, presented a case of PONHL in a 52-year-old patient [15]. Ray *et al*., in 2008, reported a case of PONHL in an 8-year-old child [16]. However, in our case, the patient presented at a very early age (1 year). In 2014, five cases of PONHL reported by Senol *et al*. presented with the complaints of fatigue, weight loss, and abdominal swelling [14]. In a study of 16 cases of PONHL by Shokralla *et al*., the majority of the patient (75%) presented with pelvic complaints, menstrual abnormalities, or constitutional symptoms [1]. Kumar *et al*., in 2014, reported a case of PONHL presented with dull aching pain for 4 months and heaviness for 2 months [17]. In the present case, the patient had acute lower abdominal pain and a lump in the right iliac fossa. We have found out no recurrence in our case over 1 year follow-up. Other studies show similar results [17,18].

CONCLUSION

PONHL is extremely rare, especially in pediatric age. The patient may present with lower abdominal pain and lump. The histopathological examination followed by immunohistochemistry can only correctly diagnose the case. Through the above case report, we wish to create awareness among the clinicians, radiologists, and pathologists to consider PONHL as a differential diagnosis of the lower abdominal mass, even in a 1-year-old child so as to ascertain the prompt diagnosis and expeditious management.

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