LETTER TO EDITOR

DOI: http://dx.doi.org/10.29387/ms.2014.2.4.147-149

Clear cell sarcoma of ovary -an exceedingly unusual neoplasm

Gupta N[1], Bharti A[2]

Correspondence to:

drnalinibansal@yahoo.com

[1]***Dr. Nalini Gupta*** MD, DNB, PDCC, Senior Histopathologist, Dept. of Histopathology, Fortis Faridabad, Haryana, India.

[2]***Dr. Ajai Bharti,*** MBBS, MS, Laproscopic Surgeon, Dept. of surgery, Jeevan Jyoti Hospital, Bareilly, Uttar Pradesh , India.

Editors for this Article:

Dr. A.K. Pradhan, MBBS, MD. Professor, KIMS, Amalapuram, Editor-In-Chief, Medical Science.

Dr. Arnab Ghosh, MBBS, MD, Professor, Pathology, MCOMS, Editorial board member, Medical Science.

Cite this article:

Gupta N, Bharti A. Clear cell sarcoma of ovary -an exceedingly unusual neoplasm. Medical Science.2014; 2(4):147-9.

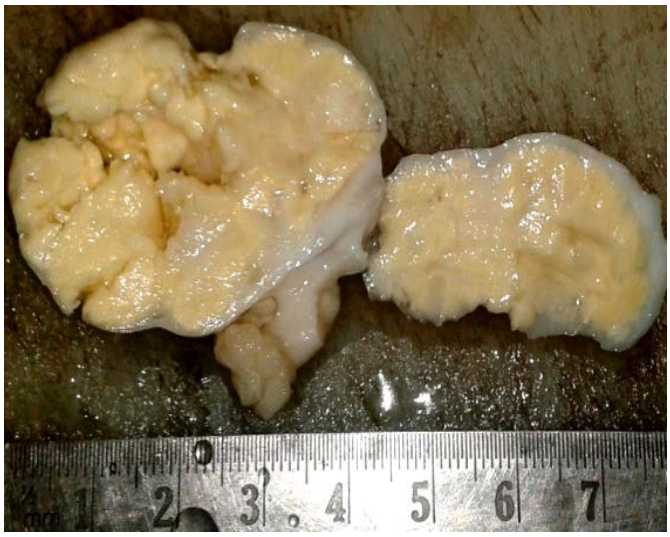
Information about the article

Received: Aug. 30, 2014
Revised: Oct. 18, 2014
Accepted: Nov. 1, 2014
Published online: Dec. 30, 2014

Dear Sir,

Clear Cell Sarcomas are uncommon neoplasm of childhood seen mostly in kidneys. Most of these tumors are aggressive with an unfavorable histology. These Tumors are mostly seen in kidneys where they have a strong propensity to metastasize to bone. Tumors with similar histology are very rarely described in other organs. There is a single case report of Clear Cell Sarcoma involving ovaries. We herein report second case of this rare entity. We report a very unusual case of Clear Cell Sarcoma of ovary in an eight month old baby girl who was admitted to Jeevan Jyoti Hospital Bareilly, with chief complaints of abdominal distention. USG examination revealed a solid right ovarian mass with moderate ascites. Her CA 125 levels were raised 274.8 u/ml. Her CEA and AFP levels were normal 0.6 ng/ml and 5.36 ng/ml respectively. Ascitic fluid cytology done was negative for malignant cells. Her other hematological investigations and Serum Creatinine, Na, K, Calcium levels were normal (0.72 mg/dl, 141mmol/L, 4.19 mmol/L and 10.0 mg/dl respectively). Based on these investigations unilateral salpingo-oophorectomy was performed. Grossly received in formalin is a right ovarian mass measuring 4 x 3 x 1.5 cms. Cut surface was solid yellowish (Figure 1).

Figure 1 - Gross showing an ovarian mass with solid grayish yellow homogeneous cut surface



ISSN 2321-5291
Medical Science 2014;2(4):147-149

Outer surface showed attached fallopian tube measuring 2 cms in length and 0.5 cms in diameter. Microscopic Examination- Sections taken from the ovarian mass show sheets and cords of clear cells having high nucleocytoplasmic ratio, hyperchromatic nuclei and moderate clear cytoplasm (Figure 2).

Figure 2 - Light microscopy showing sheets of clear cells with round hyperchromatic nuclei (H & E 40X).

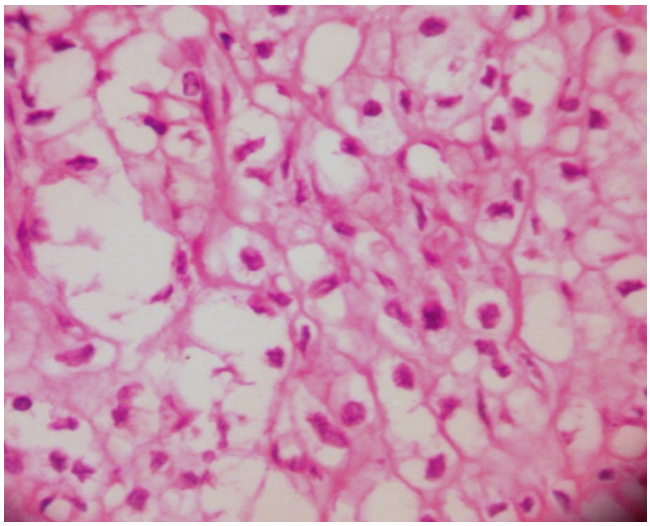
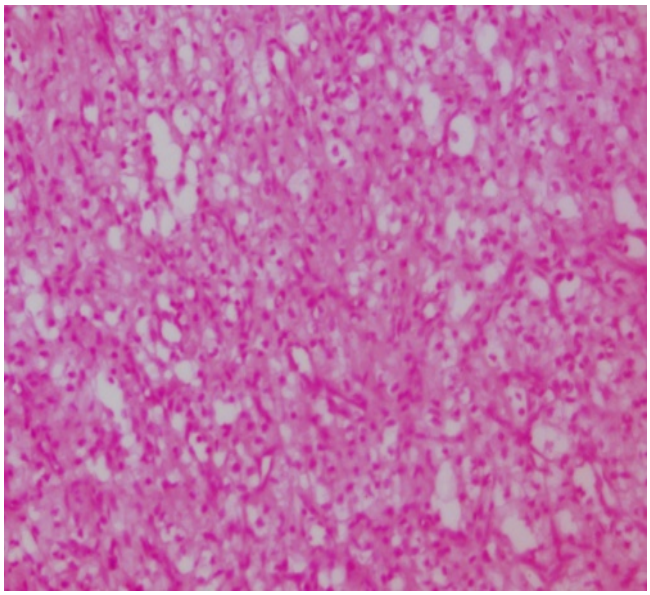
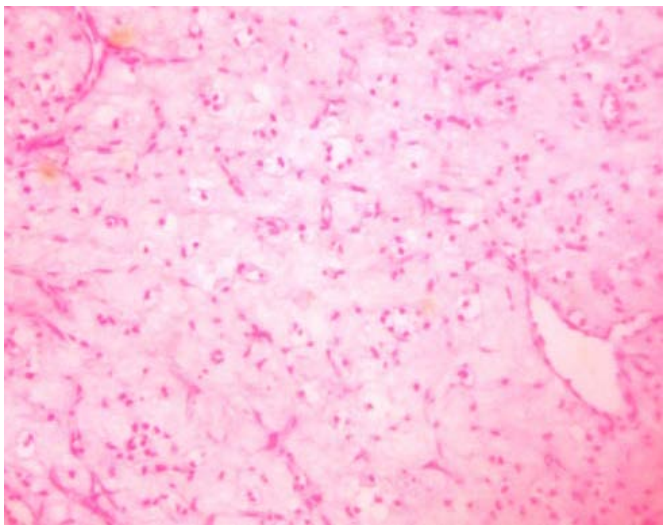


Figure 3 - Light microscopy showing clear cells with a prominent arborizing capillary network (H & E 40X).



Tumor has a prominent arborizing vascular network (Figure 3). Mucopolysaccharide matrix material is noted in between the tumor cells at places (Figure 4). No spindling of cells noted. PAS stain done is negative. IHC done for Viamentin was positive in tumor cells. Cytokeratin stain done is negative Based on these histomorphological and immunohistochemical features a final diagnosis of Clear Cell Sarcoma of ovary was made.

Figure 4 - Light microscopy showing myxomatous areas amidst tumor cells (H & E 10X).



Literature review showed only a single case of clear cell sarcoma of ovary in an infant reported by Finn et al in 2000. Since than no case with similar morphology has been reported in indexed literature. Clear cell sarcoma is very uncommon primary malignant neoplasm of childhood. They are most commonly seen in kidney where they comprise 4% of all primary renal tumors [1]. Very rarely clear cell sarcomas are reported in other organs. Only one case of clear cell sarcoma like tumor has been reported in the ovary [2]. The cell of origin of clear cell sarcoma remains uncertain till date. A study by Cutcliffe et al have postulated primitive renal mesenchymal cell that possess neural marker as cell of origin for these tumors [3]. The exact histogenesis of these tumors is not very well known. However, similar primitive mesenchymal cells may have been responsible for origin of these tumors in ovary. Grossly most of these tumors are sharply outlined with a homogeneous light gray brown color. Areas of necrosis and cyst formation can be seen [4]. Microscopically these tumors had a typical histological pattern composed of sheets of small cells with round hyperchromatic nuclei, inconspicuous nucleoli and light staining cytoplasm with indistinct cell margins set within a delicate capillary network [5]. We observed that, tumor cells are positive for viamentin and negative for cytokeratin as reported usually. Surgical intervention is required guided by Chemotherapy and radiotherapy for the treatment of Clear Cell Sarcoma. Long term follow up is essential because of late relapse. There is only a single case of Clear Cell Sarcoma of ovary reported so far in indexed literature. Histopathological features together with findings are essential for establishing a

ISSN 2321-5291
Medical Science 2014;2(4):147-149

correct diagnosis. Immunohistochemical and Histopathological features are essential for the proper diagnosis. More cases needs to be studied to determine unifying terminologies and hence the need to publish.

Key words

Clear cell Sarcoma, ovary, kidney.

Competing interests

Nil

Authors' information

Dr. Nalini Gupta, MD, DNB, PDCC, Senior Histopathologist, Dept. of Histopathology, Fortis Faridabad, Haryana, India.

Dr. Ajai Bharti, MBBS, MS, Laproscopic Surgeon, Dept. of surgery, Jeevan Jyoti Hospital, Bareilly, Uttar Pradesh , India

References

1. Beckwith JB, Palmer NF. Histopathology and prognosis of Wilms Tumors: results from the First National Wilims' Tumor Study. Cancer 1978;41(5):1937-48.
2. Finn LS, Patterson K. Ovarian sarcoma with pathologic features of clear cell sarcoma of the kidney. Pediatr Dev Pathol 2000;3(5):487-91.
3. Cutcliffe C, Kersey D, Huang CC, Zeng Y, Walterhouse D, Perlman EJ. Clear cell sarcoma of kidney: up-regulation of neural markers with activation of the sonic hedgehog and Akt pathways. Clin Can Res 2005;11(22):7986-94.
4. Sharma SC, Menon PA. Clear cell sarcoma of the kidney. J Postgrad Med 2001;47 (3):206-7.
5. Nwebould MJ, Kelsey AM. Clear cell sarcoma of the kidney in a 4 month old infant: a case report. Med Pediatr Oncol 1993; 21(7):525-8.