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Extraskeletal Primary Ewing's Sarcoma of Nasopharyngeal Region - Rare Tumour A Case Report

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***Abstract*: Ewing's sarcoma and primitive neuroectodermal tumor are closely related family of small round cell tumors seen in childhood and adolescence. The incidence of these tumors occurring in the head and neck region is just 2-7%. Ewing's sarcoma of the sinonasal tract is rare and may arise in the maxilla, and on exceedingly rare occasions in the soft tissue of the nose, i.e. as an extraskeletal Ewing's sarcoma. We report a case of extraskeletal Ewing's sarcoma of the nasopharyngeal region in a 21-year-old male who presented with nasal obstruction and epistaxis. In our case, diagnosis was confirmed by immunohistochemistry where round tumor cells were positive for CD 99. Ewing's sarcoma of nasopharyngeal region is a rare and aggressive tumor. Hence early diagnosis, combined chemotherapy, surgery and radiotherapy and long term follow up is suggested in such cases.**

***Keywords*: Ewing's sarcoma, Extraskeletal, Nasopharyngeal region, small round cell tumor.**

I. INTRODUCTION

Extaskeletal Ewing's sarcoma first described by Tefft et al., is a very rare, rapidly growing malignant round cell tumor. Ewing's sarcoma is a highly malignant small round cell tumor having both skeletal and extraskeletal forms.[1,2,3] The skeletal forms are more frequent and occur in long bones of the extremities. The extraskeletal form usually occur in the soft tissue of lower extremities, paravertebral tissues, chest wall, retroperitonium and rarely in the head and neck region.[4,5] Extraskeletal Ewing's sarcoma occurring in the Nasopharyngeal region is very rare and only few case reports have been published in literature. We report a case of Ewing's sarcoma in nasopharyngeal region in 21 year old male who is present with epistaxis and nasal obstruction. Diagnosis is confirmed by immunohistochemistry.

II. CASE REPORT(S)

A 21-year-old male visited the ENT department of Sardar Patel Medcal College, Bikaner for nasal obstruction, epistaxis and intermittent rhinorrhea for 4 months. Nasal examination revealed a mass visible in the right nasal cavity extending to the right side of the nasopharynx.

At the endoscopic examination, a lobulated mass coated with discharge was detected in the right nasal cavity expanding to contralateral side.

CT scan showed a soft tissue heterogeneous density lesion measuring 8x4.5x3 cm in the right nasopharynx involving adjacent soft tissue and destruction bone.

Fine needle aspiration was done from the nasopharyngeal mass and smears obtained were richly cellular showing loosely cohesive as well as dispersed uniform, small round cells with scant cytoplasm, and indistinct cell borders. Nuclei were

round having fine nuclear chromatin and inconspicuous nucleoli. Occasional pseudorosette formation was also seen. On the FNAC a cytological diagnosis of small round cell tumor was being kept and biopsy was advised.

After this routine investigation of patient was done. On routine investigation hematological parameter are Hemogbin 13.6%, Total WBC count – 7400cell/mm^3, Differential count: Neutrophils-76%, Lymphocytes-22, Eosinophils-01, Monocytes-01%, Platelet-178000, ESR-18mmat the end of one hour, Bleeding time-2 minute 20 seconds, Clotting time: 5 minutes 50 seconds, and HIV, HbsAg,VDRL was found to be negative by ELISA.

Urine microscopy and biochemistry was normal.

Serum electrolytes and blood sugar was in normal range.

An open surgery was done and wide excision was performed under general anaesthesia. According to operative findings, a mass in the right nasal cavity with obstruction of the left choana was noted. Destruction of posterior portion of nasal septum was also detected.

Grossly, the specimens were irregularly fragmented myxoid and necrotic mucosal tissues measuring 10x5x4cm and cut surface is gray white.

Histopathology- Multiple section was taken and stained with hematoxylin and eosin and examine under microscope. The tumor was composed of densely distributed, uniform, small- to medium-sized, round cells with scanty cytoplasm. The cells showed mild anisonucleosis with round to oval nucleus having fine granular chromatin with 0-1 inconspicuous nucleoli (figure1, 2, 3). In addition, the tumor cells also showed strong positivity for PAS stain (figure 4). On the basis of histopathological examination differential diagnosis of Ewing's sarcoma, PNET, neuroblastoma or rhabdomyosarcoma were suggested and immunohistochemical panel for vimentin, desmin, chromogranin and CD99 was put. The present case showed strong positivity for CD99 (figure 5) but was however negative for desmin and chromogranin.

On the basis of histological, immunohistochemical, clinical and radiological findings a final diagnosis of Extraskeletal Ewing's sarcoma of nasopharyngeal cavity was made. Following the diagnosis, the patient was immediately put on chemotherapy as Extraskeletal Ewing's sarcoma is a radiosensitive tumor. After 2 cycles of chemotherapy the tumor size decreased significantly and the patient responded very well to therapy with improvement in her general condition.

III. DISCUSSION

There has been remarkable evolution in the concepts regarding Ewing's Sarcoma histogenesis and relation with other small round cell tumors, including peripheral primitive neuroectodermal tumor (pPNET). The relationship between Extraskeletal Primary Ewing's Sarcoma and pPNET represents one of the most fascinating controversies in pathology. Both Extraskeletal Ewing's Sarcoma and pPNET show varying degree of neuroectodermal differentiation.[6]

The term Extraskeletal Ewing's Sarcoma has been used for those tumors that lack neuroectodermal differentiation by light microscopy, immunohistochemistry and electron microscopy.[7]

Extraskeletal Ewing's Sarcoma arising in head and neck is extremely rare and only some cases have been reported in the orbit, scalp, face, nasal cavity, paranasal sinus, nasopharynx, parapharyngeal space, larynx, hard palate, submandibular gland, parotid gland, thyroid gland, and soft tissue of the neck.

CT and MRI are frequently used in the radiological evaluation of Extraskeletal Ewing's Sarcoma of the head and neck. However, these modalities cannot provide a specific diagnosis, but successfully demonstrate the internal structure of the lesion and the extent of the tumor.

Ewing's sarcoma is distinguished by two types: skeletal type and extraskeletal type. Most commonly, Ewing's sarcoma arises from skeletal structures, especially long bones (35%), and pelvis (24%). The extraskeletal type of Ewing's sarcoma usually occurs in the soft tissue of the lower extremities and the paravertebral region.

Thirteen cases of primary Ewing's sarcoma in the nasal cavity and/or paranasal sinuses have been reported in the otolaryngology literature. The prognosis for Ewing's sarcoma depends on the presence of metastases, because Ewing's sarcoma is highly malignant and metastasizes early to bones and lungs. Recently, it is accepted that prompt chemotherapy is necessary to treat occult metastasis, and a combination of surgical excision, radiotherapy and chemotherapy has significantly improved the 5-year survival rates, now reaching to 75%.[8,9]

Microscopically, Ewing's sarcoma shows uniform, small, round cells with round to elongated nuclei, scanty cytoplasms and indistinct cytoplasmic borders. Hemorrhagic areas and extensive necrotic lesions are common. The essential diagnostic examination for Ewing's sarcoma among many small round neoplasms is CD99 marker, the specific immunohistochemical examination. In addition, molecular studies using PCR to detect characteristics of chromosomal translocations are definitive for the diagnosis of Ewing's sarcoma. Specific genetic hallmarks of Ewing's sarcoma is a gene sequence t(11;22)(q24;q12), which results in the fusion of the EWS gene with the FLI gene.[10]

Ewing's sarcoma is composed of uniform small, round, undifferentiated tumour cells with round or oval nuclei exhibiting a fine chromatin pattern, small nucleoli and scanty cytoplasm usually crowded in sheets or segregated in lobules by fine fibrovascular septa.[10] The intracytoplasmic glycogen may be demonstrated by PAS stain in 75% of the cases, but it is not pathognomonic and conclusive because other small round cells may show the presence of glycogen as well. Since Ewing's sarcomas are usually vascular; hemorrhagic areas and extensive necrosis are common. Histopathologically the tumor must also be differentiated from other small round cell tumor such as mesenchymal chondrosarcoma, rhabdomyosarcoma, malignant lymphoma, eosinophilic granuloma, neuro-endocrine tumors and metastatic neuroblastoma.[10,11] The use of immunohistochemistry has helped in the diagnosis of this tumor. In general, the tumor cells are positive for vimentin and CD99 and negative for neural, skeletal, vascular and lymphoid cell markers.[12,13,14] Regarding Mic-2 antigen, recently published data have confirmed the high sensitivity of the Mic-2 gene product (CD99) for all Ewing's Sarcoma family tumors with over 95% of the cases showing positivity for this marker.[12,13]

IV. CONCLUSION

We hereby conclude that although wide spectrum of small round cell tumors cause significant diagnostic challenges, careful evaluation and sound knowledge of light microscopic features with the help of ancillary techniques like immunohistochemistry and cytogenetics will help for accurate diagnosis and appropriate management of the patient of Ewing's sarcoma.

Conflict of interest - none

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Competing Interests

- Competing interests: None declared

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APPENDIX - A

FIGURE WITH LEGENDS

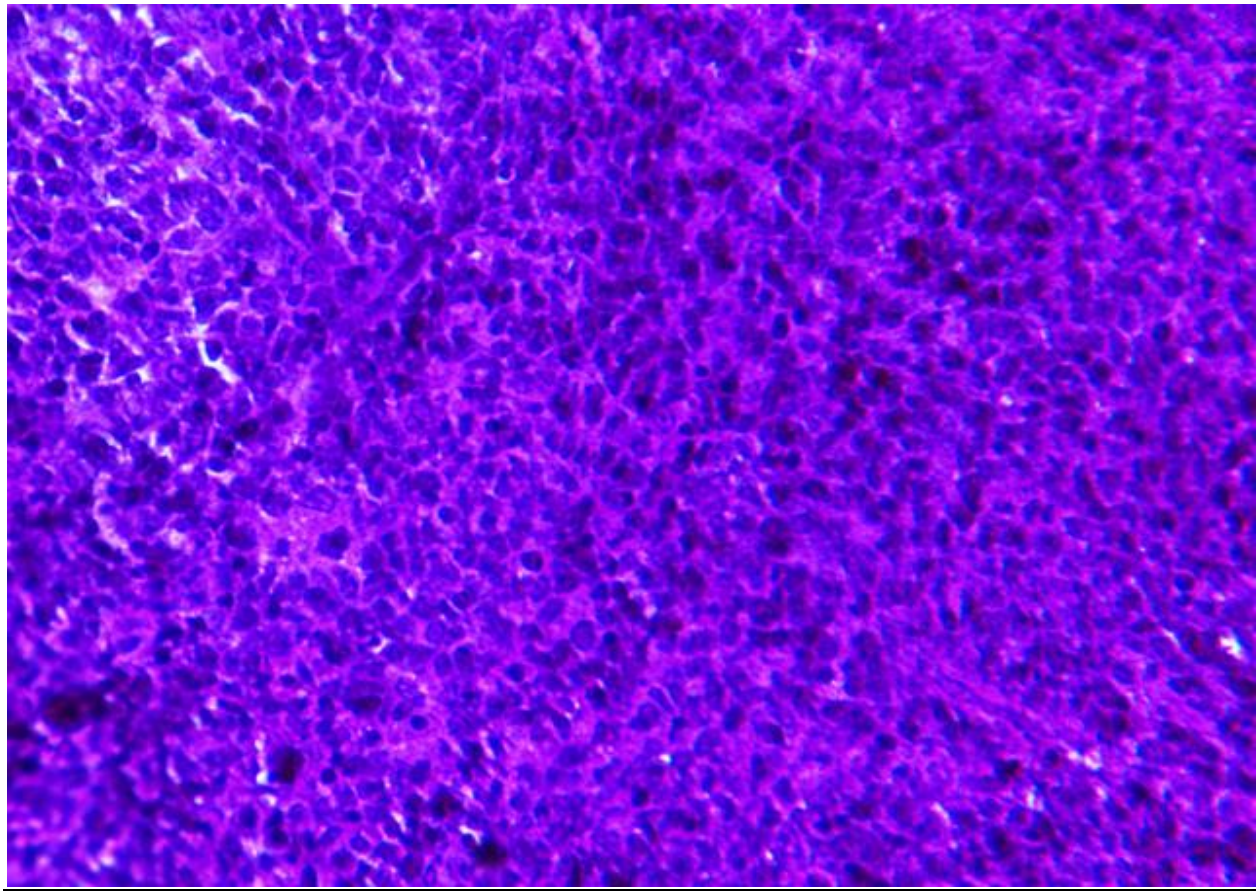

Figure 1- section shows uniform small round cell with scanty eosinophilic cytoplasm, indistinct cytoplasmic borders and minimal pleomorphism. (high power)

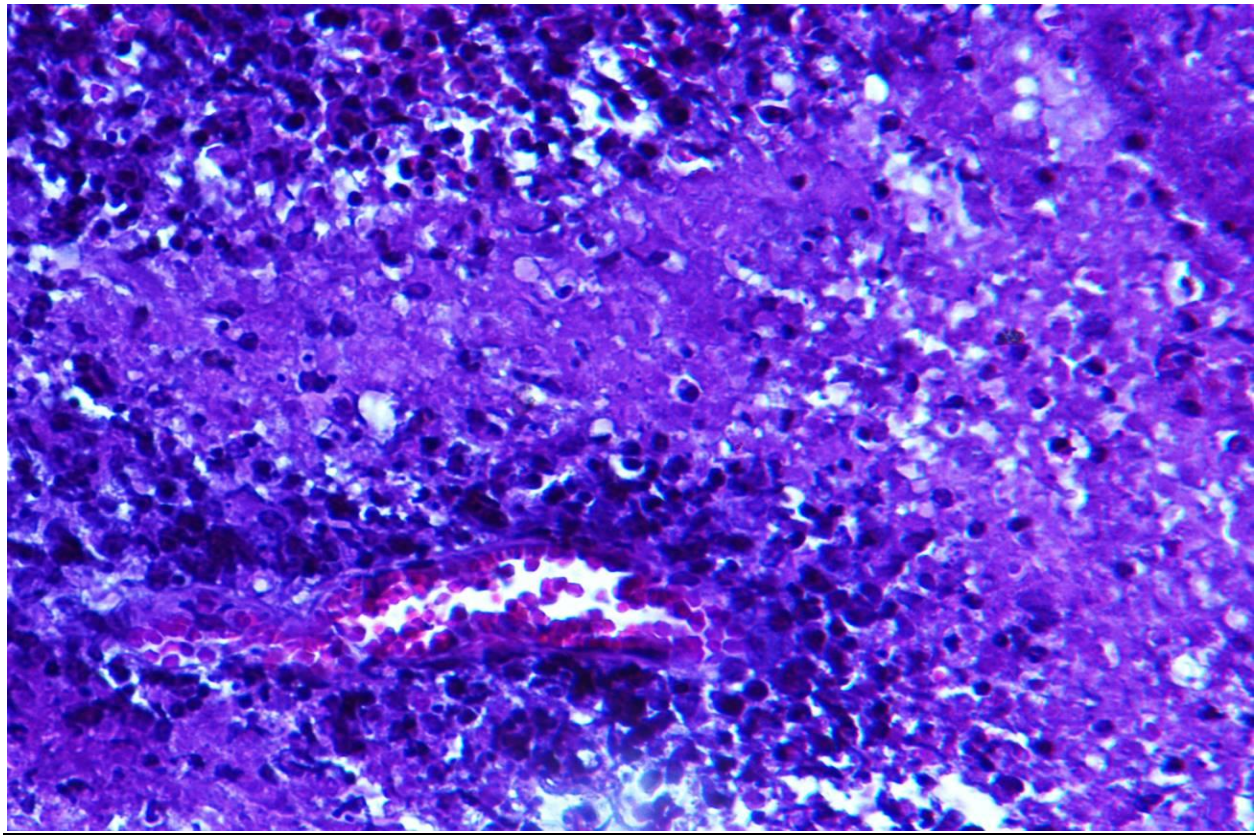

Figure 2- Tumor cells are uniform round with minimal cytoplasm and surround the blood vessel forming pseudorossette. (high power)

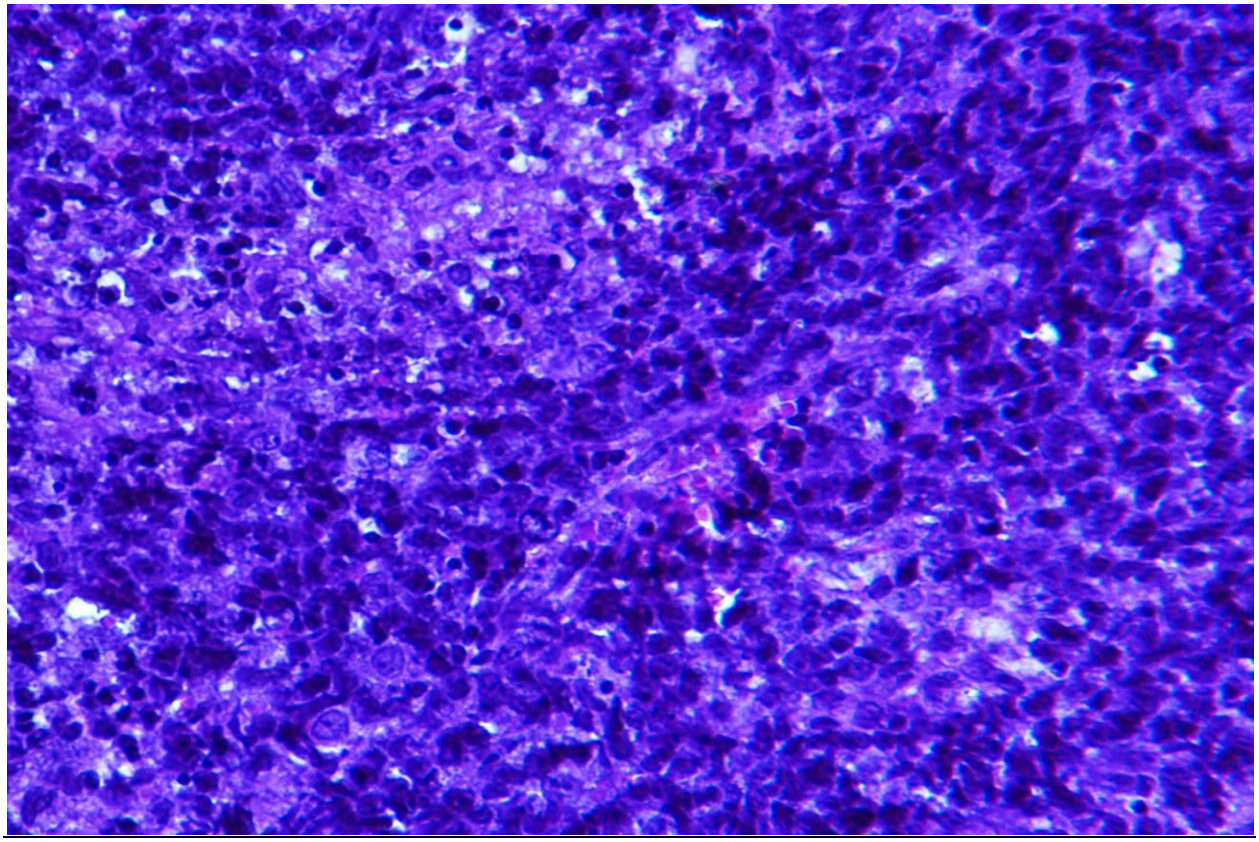

Figure 3- Tumor cells are uniform small round cells arranged in loosely discohesive pattern and having scanty cytoplasm. (high power)

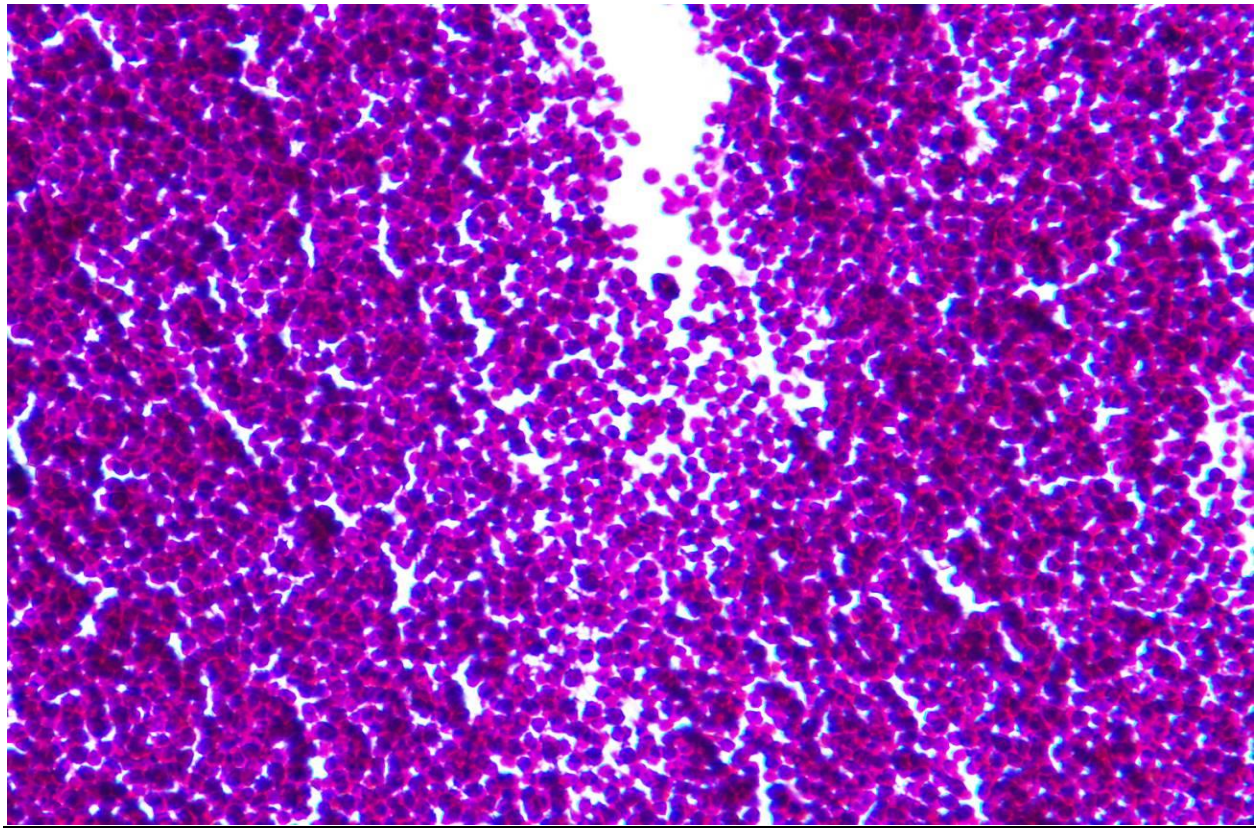

Figure 4 – shows PAS staining of tumor which demonstrate intracytoplasmic glycogen stain by PAS staining and give magenta colour to cytoplasm.(40X)

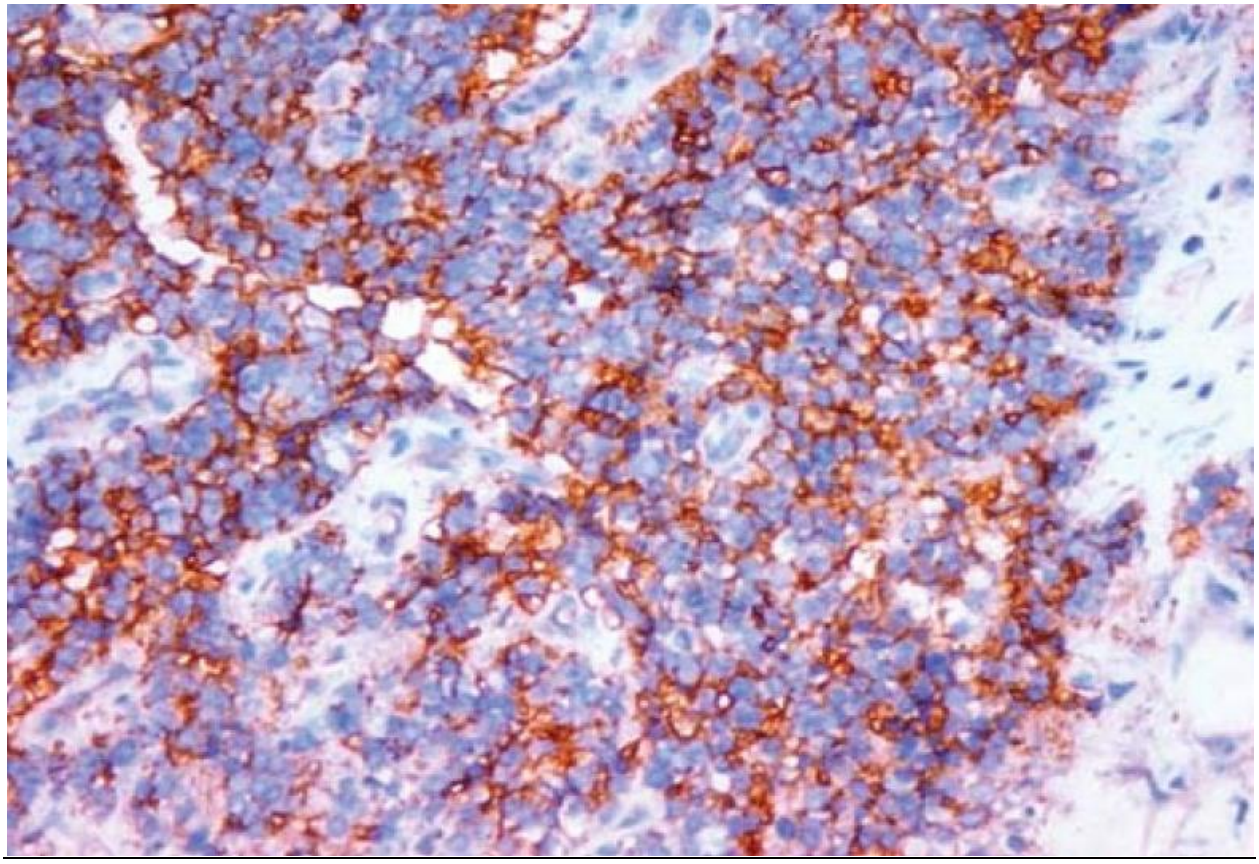

Figure 5– shows Immunopositivity for CD99 which is diffusely positive for CD99.(Low Power)