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Round Cell Lesion

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Abstract

Round cell tumors as the name suggest are comprised round cells with increased nuclear-cytoplasmic ratio. This group of tumor includes entities such as Ewing sarcoma,Wilms tumor/nephroblastoma,neuroblastoma,desmoplastic small round cell tumor. These round cells tumors are characterized by typical histological pattern, immunohistochemical and electron microscopic features that can help in differential diagnosis. The present article describes the classification and explains the histopathology and radiological features of some important round cell tumors.

Keywords: Ewing sarcoma, Wilm's tumor, neuroblastoma, desmoplastic small round cell tumor.

Introduction:

Round cells are united by morphology and differs greatly in Histogenesis. It is generally consist of sheets of round cell which are small having large nucleus, scanty cytoplasm and a hyper-chromatic or vesicular nucleus. They have highly undifferentiated neoplasms composed of a monotonous population of round cells with high Nuclear Cytoplasm ratio.

Histological similarity, undifferentiated and primitive character makes diagnosis difficult, especially the tumors are poorly differentiated. The possibility of definitive diagnosis seems likely. Therefore, light microscopic histology remains as gold standard in diagnosis, followed by immunohistochemical evaluation [1].

Malignant small round cell tumors (MSRCT) is a term used for tumors composed of malignant round cells that are slightly larger or twice the size of red blood cells in air-dried smear [2].

Round cell neoplasm – heterogeneous group of neoplasm characterized by the sheets of poorly differentiated cells.

- Small [similar to lymphocyte in size]
- Round [round nuclei and scanty cytoplasm]
- Blue [due to high nuclear cytoplasmic ratio]

Epidemiology:

The rarity of Desmoplastic small round cell tumor (DSRCT) with an incidence of 0.2–0.5 cases/million. Hinders data acquisition and analysis of patient characteristics. Recently, a retrospective studies and database analysis evaluated international populations with 50–491 patients. DSRCT affect primarily adolescents and young adults with high incidence between the age of 20 and 25 years. DSRCT shows up as a striking man.

Etiology:

The causes for round cell tumor is unknown . Cancer begins when cell develops alteration in its DNA. A cell DNA contains the instructions the cell what to do. This change causes rapid multiplication of cell. This creates a cluster of cancer cells called a tumor. Cancer cells invade and destroy body tissue. Over time, cancer cells are separated away and spread to other parts of the body[3].

Classification:

1.According to the size of round cell:

- Round cells were also based on size, often seen in Squamous cell carcinoma, Ewing's sarcoma, melanoma, Rhabdomyosarcoma (RMS), Langerhans cell disease, lymphoma, adenocarcinoma, neuroendocrine carcinoma, Merkel cell carcinoma and olfactory neuroblastoma.
- Large round cell type can be found in Squamous cell carcinoma, adenocarcinoma, melanoma, RMS, lymphoid tumors, paraganglioma.

2. Basis on origin:

- Neurogenic origin: Ewing's sarcoma, neuroblastoma, retinoblastoma, medulloblastoma, Merkel cell tumor, paragangliomas, small cell lung tumor Mesenchymal in origin.
- Myogenic differentiation: ERMS, ARMS.
- Osteoid differentiation: Small cell osteosarcoma.
- Chondroid differentiation : Mesenchymal chondrosarcoma.
- Adipose tissue like differentiation can be seen in myxoid/round cell liposarcoma. Lymphoma is haematolymphoid in origin.
- Malignant soft tissue tumors of uncertain type: DSRCT and poorly differentiated synovial sarcoma[4].

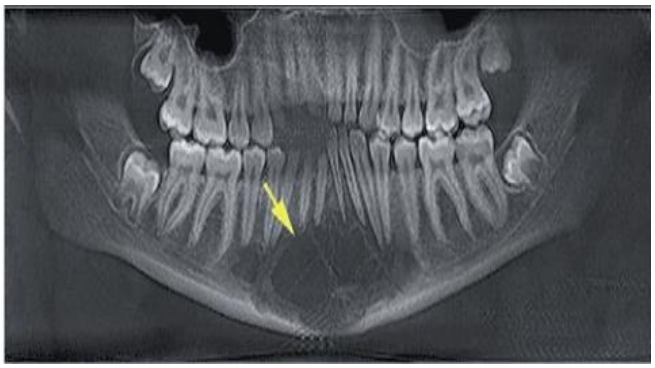
1.EWING'S SARCOMA / PRIMITIVE NEUROECTODERMAL TUMOR:

- *CLINICAL FEATURES:*

The first clinical manifestation was swelling and followed by swelling and pain. Dental displacement, neck adenopathies , fever, dental mobility, root resorption, destruction of the dental follicle and premature dental exfoliation. Ewing sarcoma was predominantly hard in consistency (29.6%) and the overlying mucosa was usually normal in appearance (15.5%).[5,6]

- *RADIOLOGICAL FEATURES:*

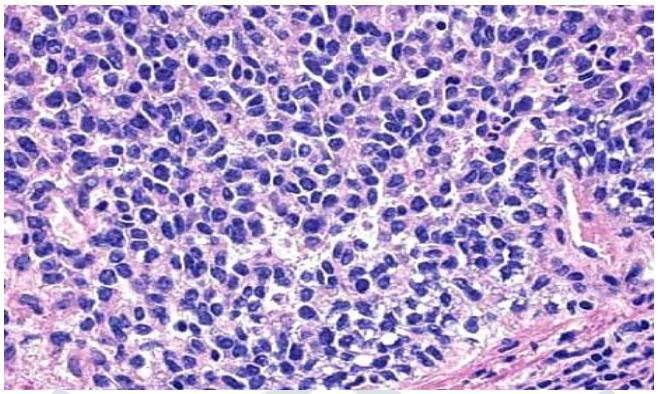
The lesion is destructive and produce an irregular, poorly defined, diffuse radiolucency, while lesions of the jaw resembles sclerosing osteomyelitis have been reported. A common characteristic radiographic feature is the formation of layers of new sub-periosteal bone producing 'onion skin' appearance on the long bone radiograph but rarely present in the jaw. This thickened cortex is normally infiltrated by tumor. Osteophyte formation may view on the radiograph and sometimes, may be similar to the 'sun-ray' appearance of osteosarcoma. The extra skeletal lesions may cause erosion, cortical thickening or osseous invasion to the adjacent bone .[4]



- *HISTOPATHOLOGICAL FEATURES:*

Ewing sarcoma is an extremely cellular neoplasm composed of solid sheets or masses of small round cells with very little stroma, although few connective tissue septa may be present. The cells are small and round, with scanty

cytoplasm with relatively large round or ovoid nuclei with dispersed chromatin and hyperchromatic . The cell borders are indistinct. Sarcoma cells are arranged in "Filigree pattern". Mitotic figures are commonly seen . The cells are optimistic for glycogen and diastase resistance. Rosettes present in 10% of cases. Many tiny vascular channels may also be seen. Haemorrhage with vascular lakes or sinuses may be seen. Geographic necrosis with perivascular sparing is a common feature. Increased cellular pleomorphism and increased numbers of bizarre giant cells may be found in the lesions in patients treated with radiation and adjuvant chemotherapy [4].



2. RHABDOMYOSARCOMA:

In 2013, WHO classifies rhabdomyosarcoma into four categories:

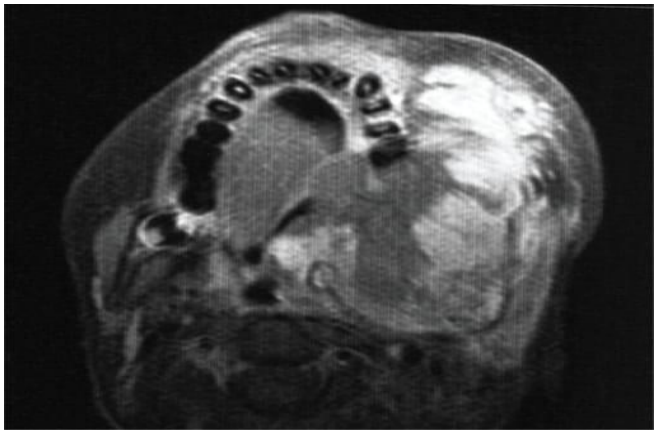
- Embryonic
- Alveolar
- Pleomorphic
- Spindle/Sclerosing

➢ *CLINICAL FEATURES:*

The most commonly affected areas are the head and neck region, genitourinary tract, retro peritoneum, and, to a lesser extent, the extremities [2]. The head and neck RMSs are anatomically divided into 2 categories: para meningeal (including RMS of the nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infra-temporal fossa and pterygopalatine fossa) and non-parameningeal (including RMS of the scalp, orbit, parotid gland, oral cavity, oropharynx and larynx). RMS of the oral cavity accounts for 10–12% of all the head and neck RMS cases. [8,9,10]

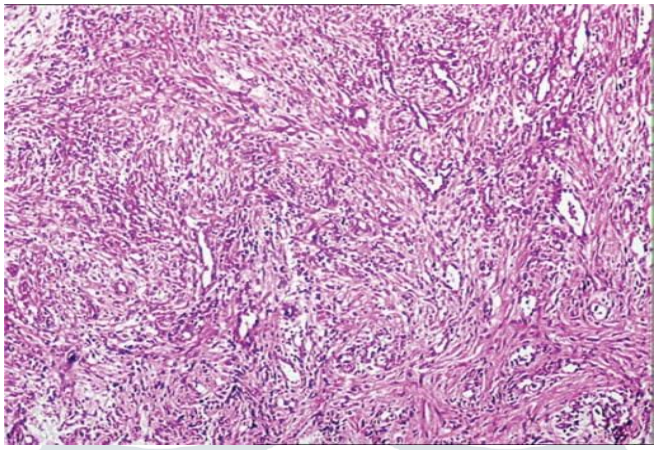
➢ *RADIOLOGICAL FEATURES:*

Magnetic resonance imaging (MRI) showed an infiltrative large soft tissue lesion in the maxillary sinus, infra orbital space, pterygopalatine fossa, pterygoid plates and the ramus, which caused displacement of the adjacent structures. [11]



- *HISTOPATHOLOGICAL FEATURES:*

Based on histological findings, 4 broad
subtypes of RMS have been identified: botryoid and spindle cell RMS, embryonal RMS, alveolar RMS, and undifferentiated RMS. The histogenesis of RMS is unknown, but the most widely accepted hypothesis is that, RMS arise due to proliferation of embryonic mesenchymal tissue. [12,13,14]



3. WILM'S TUMORS / NEPHROBLASTOMA:

- *CLINICAL FEATURES-:*

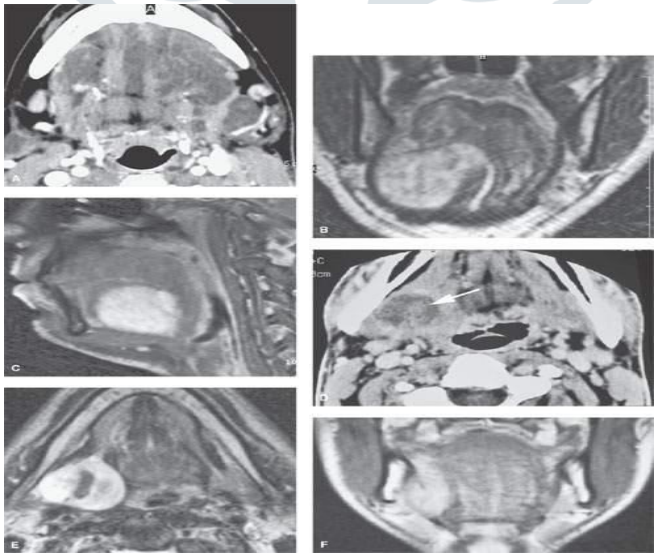
Wilm's tumor is unrelated to adult kidney cancer. The tumor usually affects a single kidney, but approximately 5-10 percentage of children with Wilms tumor involve both kidneys. A mass in the stomach is felt, Swelling present in the stomach area. Pain in the stomach region. May include Fever, Blood in the urine, Low red blood cell level(anaemia), High blood pressure [15].

- *RADIOLOGICAL FEATURES:*

Abdominal x-ray typically reveals a large soft tissue opacity displacing bowel.

It appears as solid mass on ultrasound that may have vascular invasion with smooth margin. Cystic or necrotic area are anechoic and hypoechoic respectively while haemorrhage/fat/calcification appear as hyperechoic.

These tumors appear heterogeneous on all sequences and frequently contain blood products in MRI Scan. [16,17]



- *HISTOPATHOLOGICAL FEATURES:*

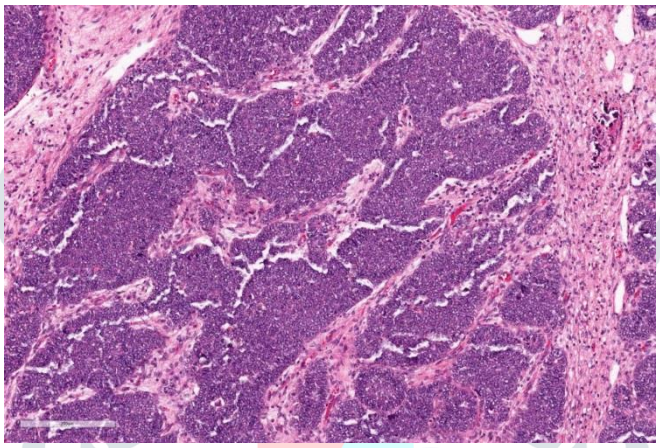
Consists of 3 components: Blastemal, Epithelial and Stromal. Tumor showing two or only one component not rare, Blastema least differentiated component consists of small to medium sized undifferentiated cells with relatively small

regular nuclei and nucleoli shows different patterns (diffuse, serpentine, nodular and basaloid patterns) which has no prognostic significance, Mitotic figures are frequently seen.

Epithelial component comprises:

- Poorly-differentiated (rosette-like structure)
- Moderately-differentiated (tubules and papillary structure)
- Well-differentiated (glomerular-like structures and small mature tubules) element may contain heterologous elements (squamous and mucinous epithelium, glial tissue)

Stromal component from hypo-cellular to hyper-cellular, undifferentiated areas to well-differentiated area. Cells showing no clear cell borders, oval to spindle shaped nuclei with bland , nucleoli Often shows heterologous elements (Rhabdomyoblast , adipose tissue, cartilage).[18]



4. NEUROBLASTOMA:

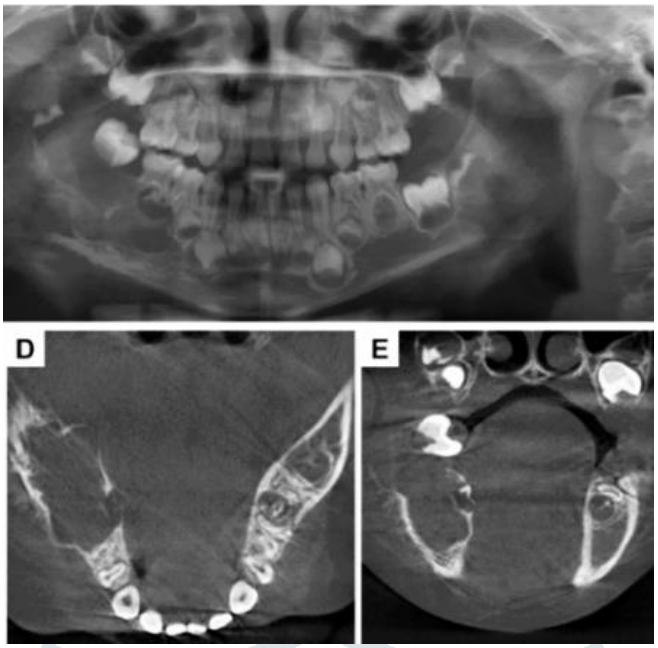
- *CLINICAL FEATURES:*

In many cases, oral metastases were found in a substantial number of patients with neuroblastoma, this condition is considered in the differential diagnosis of jaw lesion by Dentist.

83 patients with neuroblastoma were examined respectively to determine the frequency with which this malignancy affects the osseous structures of the oral cavity. These structures were involved in 21 (25%) of all patients and in 21 (49%) of the 43 with bone metastasis as judged from clinical, radiologic, and pathologic findings. Evidence of oral involvement lead to diagnosis in two instances and was discovered during the initial evaluation in 10 of the 21 patients with facial manifestations. [19]

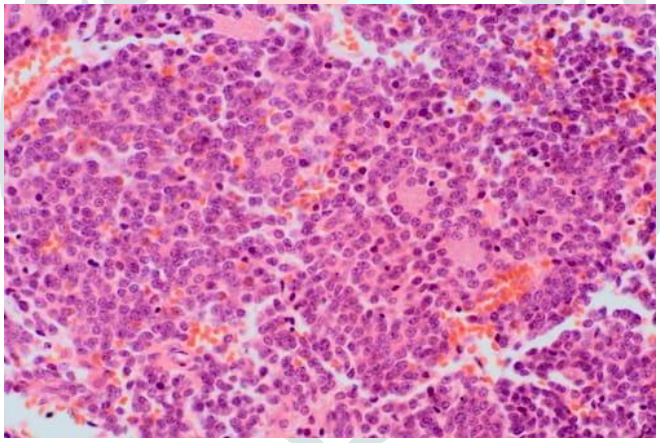
- *RADIOLOGICAL FEATURES:*

The most important radiologic findings were lytic lesions, expansion of the dental follicle and poorly defined borders around the crypts of developing teeth. [19]



- *HISTOPATHOLOGICAL FEATURES:*

Neuroblastoma is one of the "small round blue cell tumor" most typically seen in childhood. These neoplasms recurrently arise in the adrenal gland; they can reach large size in the retro-peritoneum before detection. They often contain areas of necrosis and calcification. The rosette formation, with cells arranged around a centre of cellular fibrillar extension. At higher magnification, the round cells with prominent blue nuclei, granular chromatin and minimal cytoplasm are observed [19].



5.DESMOPLASTIC ROUND CELL TUMOR:

- *CLINICAL FEATURES:*

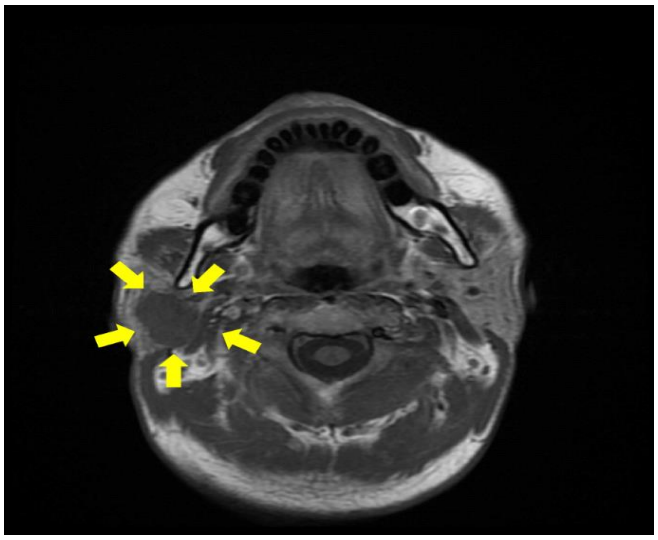
DSRCT symptoms vary depend on where the cancer initiated. Most often it initiated in the abdomen.

Signs and symptoms of desmoplastic small round cell tumors in the abdomen contains:

Swelling of the abdomen, Abdominal pain, Constipation, Difficulty urinating. [3]

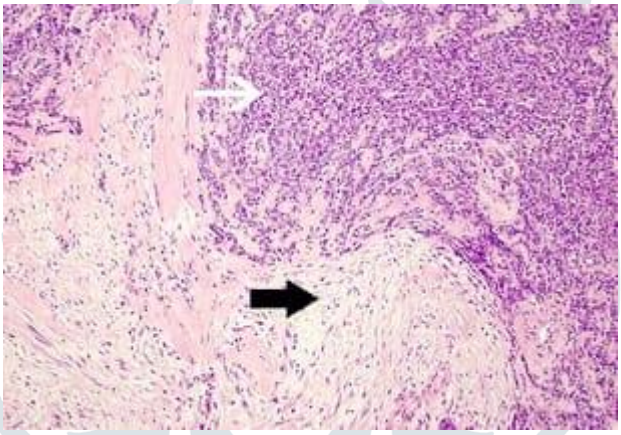
- *RADIOLOGICAL FEATURES:*

CT: Solitary or multiple soft tissue mass are seen with no definite organ of origin, usually in the retrovesical or rectouterine space, which improves heterogeneously on contrast studies. Necrosis, haemorrhage and fibrous components are common.Commonly spreads via Peritoneal seeding, lymph nodal involvement, liver and bone metastases. [20,21]



- *HISTOPATHOLOGICAL FEATURES:*

It is round, oval cells with uniform morphology embedded in an abundant desmoplastic stroma. Tumor cells were arranged in small clumps, large nests, trabecular or cord-like pattern, possessing hyperchromatic nuclei with unclear nucleoli and pale eosinophilic cytoplasm with indistinct margin. The mitotic figures were easily identified. Desmoplastic stroma was composed of spindle cells mimicking fibroblasts/ myofibroblast, accompanied by hyaline degeneration and vascular proliferation in some areas. [4]



DIAGNOSIS:

Biopsy:

•Bone marrow biopsy

• needle biopsy

• surgical biopsy

Imaging:

• bone X-ray

• MRI (Magnetic Resonance Imaging)Scan

• CT (Computed Tomography)Scan

• chest X-ray

• bone scan

• positron emission tomography (PET) Scan

Laboratory test:

- Testing of the tumor via biopsy sample and blood tests, which may aid other diagnostic tools.
- Immunohistochemistry
- FNAC (Fine Needle Aspiration Cytology)
- Reverse transcriptase PCR (RT-PCR)

Medical history and physical examination.

TREATMENT :

There is no standard treatment for recurrent Ewing sarcoma but treatment options may include the following: Combination chemotherapy, Surgery, Radiation therapy to bone tumors, as palliative therapy to relieve symptoms and improve the quality of life**.**

CONCLUSION :

The round cell tumors depict a group of highly aggressive and undifferentiated malignant group of tumors consisting of a monotonous population of round cells. The prevalent distribution and heterogeneous histology of different round-cell tumors creates difficulty in diagnosis. Hence, round cell tumors certainly represent the best model to how the integration of different diagnostic tools (morphology, immuno-histochemistry, and molecular genetics) may impact on the accurate diagnosis of tumors.

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