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RESEARCH ARTICLE

A RARE CASE OF LARGE SIZE UNDIFFERENTIATED PLEOMORPHIC SARCOMA OF ANKLE

Dr. Deeravathu Karthik Naik[1] and Dr. Adiraju Kalyani[2]

1. Post Graduate, Department OG General Surgery Siddhartha Medical College, Vijayawada, Andhra Pradesh.
2. Assosciate Professor, Department OG General Surgery Siddhartha Medical College, Vijayawada, Andhra Pradesh.

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Abstract

Undifferentiated sarcoma shows no identifiable line of differentiationwhen analyzed by presently available technology (WHO 5th edition). Occurs as 20% of all soft tissue sarcomas.tumor most frequently occurs in soft tissues of extremeties and retroperitoneum .Peaks around age of 60 years .More common in men.It begins as a painless enlarging mass ,generally in relation to skeletal muscle,deep fascia or subcutaneous tissue.MRI is the investigation of choice for all soft tissue sarcomas.Prognosis include depth of tumor and staging of tumor .Surgerical resection of tumor fallowed by radiotherapy is main stay of treatment.

Introduction:-

Definition:-
Undifferentiated sarcoma shows no identifiable line of differentiation when analyzed by presently available technology (WHO 5th edition).

Epidemiology:-
Accounts for as many as 20% of all soft tissue sarcomas.Tumor occurs most frequently in the soft tissues of extremities (50% in lower limbs, 20% in upper limbs) and occasionally in bone and viscera (WHO 5th edition).Peaks around the age of and is more common in men.

Pathophysiology:-
Genetic alterations, such as mutations, deletions, epigenetic modifications, may be important for undifferentiated sarcoma development and progression; although they are nonspecific.Recent investigation had classified TP53, ATRX, H3F3A, ZFHX3, CSMD3, PRPRT, TRIO, CLTC, PDGFRB, ALK, PTCH1, RET, ERBB4, JAK3, GATA1, PIK3CG, RARA and MYH9 as cancer driver genes

Investigations –
MRI is the investigation of choice for soft tissue sarcomas.In settings where MRI is not feasible CT can also be done.

Prognostic Factors :-
Deep tumor location and AJCC stage are the most important predictive prognostic factors .Other factors that indicate a poorer prognosis include inadequate surgical margin around the tumor, metastatic spread, large tumor size and

Corresponding Author:- Dr. Deeravathu Karthik Naik
Address:- Post Graduate, Department OG General Surgery Siddhartha Medical College, Vijayawada, Andhra Pradesh.

older age.Vast majority of undifferentiated sarcomas are high grade lesions, with a local recurrence rate of 19 - 31%, a metastatic rate of 31 - 35% and a 5 year survival of 65 - 70%

Treatment :
Surgery in combination with radiotherapy (sporadically combined with chemotherapy in case of high risk of development of metastasis) is the common therapy of choice for undifferentiated sarcoma, which is similar to the treatment of other soft tissue sarcomas

Case Report-
A 78 Year old patient presented with complains of nodular growth on anterior aspect left ankle for about 6 months, started as a small nodule of size 3*2 cm and progressed rapidly as ulcero proliferative growth to present size

History of decreased appetite and significant weight loss present from past three months.No history of comorbidities.

Clinical Examination –
Patient has a solitary 10*10*2 cm irregular shaped ulcero proliferative growth extending along the lateral and posterior aspect of left distal leg,ankle and foot region with well defined and irregular margins,lobulated surface, surrounding skin appears normal (Fig A ,B, C).Swelling is firm in consistency with no local rise in temperature and tenderness, non mobile and fixed to skin on palpation. Left superficial inguinal lymph nodes were palpable which are 2*1 cm size, non tender, discrete, hard in consistency ,mobile.
His vitals are stable

Radiological findings-
On Ct Left Ankle And Foot-
14.8*11.7*3.2 cms well defined lobulated soft tissue density mass lesion is noted along the lateral aspect of distal leg,ankle and foot region.Lesion is seen involving the skin and subcutaneous planes.no calcifications are noted.Minimal peri osteal reaction is noted along distal shaft of tibia and fibula.NO Bone destruction is noted.No sclerotic lesions noted

Chest x ray,HRCT Chest ,CT Abdomen,CT Brain and whole spine shows no evidence of metastasis

LAB-

HEMOGLOBIN(g/dl)	**5.7**	11.6	14.8
LEUCOCYTE COUNT(*1o3µ/L)	**45.6**	3.3	8.6
NEUTROPHIL(%)	**95**	38	77
PLT (*104 /µL)	17	15.8	34.8
CRP(mg/L)	59.1		
Alb(g/dl)	3.0	4.1	5.1
AST(U/L)	25	13	30
ALT(U/L)	10	7	23

Patient had normocytic normochromic Anemia with Neutrophilic leukocytosis and rest of the blood parameters are normal

Biopsy Report:-
Sections showed tumor comprised of oval to spindle cells arranged in fascicle and in bundles along with myxoid patternand increased vascularity .cells show marked pleomorphism and increased mitotic activity -10-12/10 HPF .Also show necrosis and severe Chronic non specific inflammatory infiltrates .(Fig D,E,F)

Impression:- Histological features suggestive of "**UNDIFFERENTIATED PLEOMORPHIC SARCOMA "**

Fnclcc Grading –
Tumor differentiation score – 3,Necrosis score – 1,Mitosis score 2

Total score 6, GRADE 3

Fnac Of Inguinal Lymph Node:-
Features suggestive of CHRONIC REACTIVE LYMPHADENITIS .NO EVIDENCE OF MALIGNANCY

On Clinical,pathological and radiological a diagnosis of UNDIFFERENTIATED PLEOMORPHIC SARCOMA was made with TNM staging **T2N0M0 WITH STAGE 3A and as tumor is of large size and** unable **to reconstruct the flaps we planned to go for BELOW KNEE AMPUTATION.**

After the initial pre operative workup and pre anesthetic assessment ,a detailed informed and written consent was obtained for Below knee amputation .

Intra Operative Findings- In supine position under spinal anasthesia left leg painted and drapped with 2% betadine .Anterior skin incision is given 12cm from tibial tuberosity,skin incision continued transversely up to 1/3rd of calf circumference,skin incision is then exteneded along the vertical axis 1.5 times length of transverse incision.Anterior compartment muscles divided at level of skin incision.Anterior pedicle (Anterior tibial artery and vein ,deep peroneal nerve)identified and then vessels are ligated and nerve is transected under tension.Tibia is cleared from lateral and posterior group of muscles with periosteal elevator .Tibia is divided 2cm proximal to skin insicion by giglisaw.Lateral compartment muscles are identified and divided to expose fibula.Fibula is then divided 2cm proximal to tibial transection.posterior group of muscles divided obliquely and posterior flap is created.posterior pedicle(Posterior tibial vessels and tibial nerve) identified and ligated.Hemostasis achieved .CRD drain place and fixed with skin.fasica is closed with 2-0 vicryl with interrupted absorbable sutures.skin is closed with 1-0 Ethilon in vertical matress.
(Fig I TO M)

Pictures:-

Figure A,B,C:- Ulcero proliferative growth on left ankle (Anterior view,Lateralview,Posterior View Respectively).

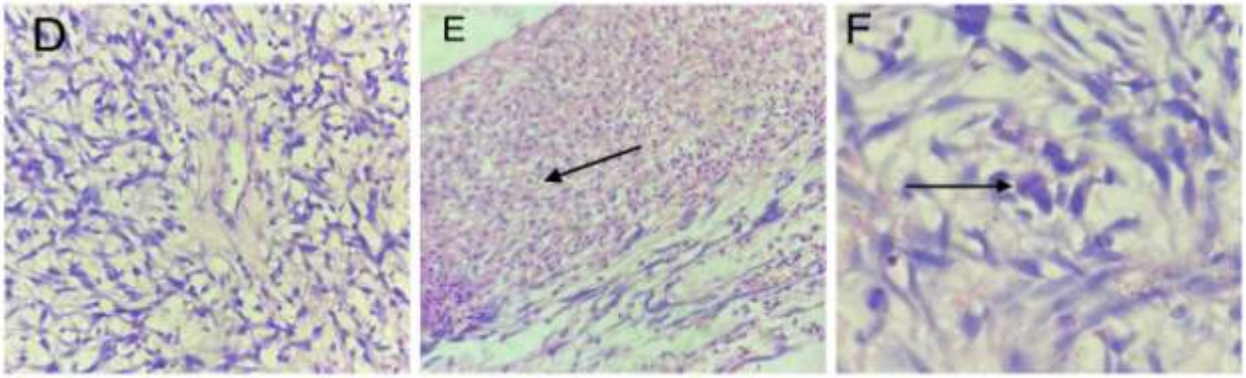

Figure D:-Histopathological examination showing oval to spindle cells with inflammatory cells
Figure E:-Histopathological examination showing necrosis (indicated with arrow).
Figure F:-Histopathological examination showing mitotic activity.

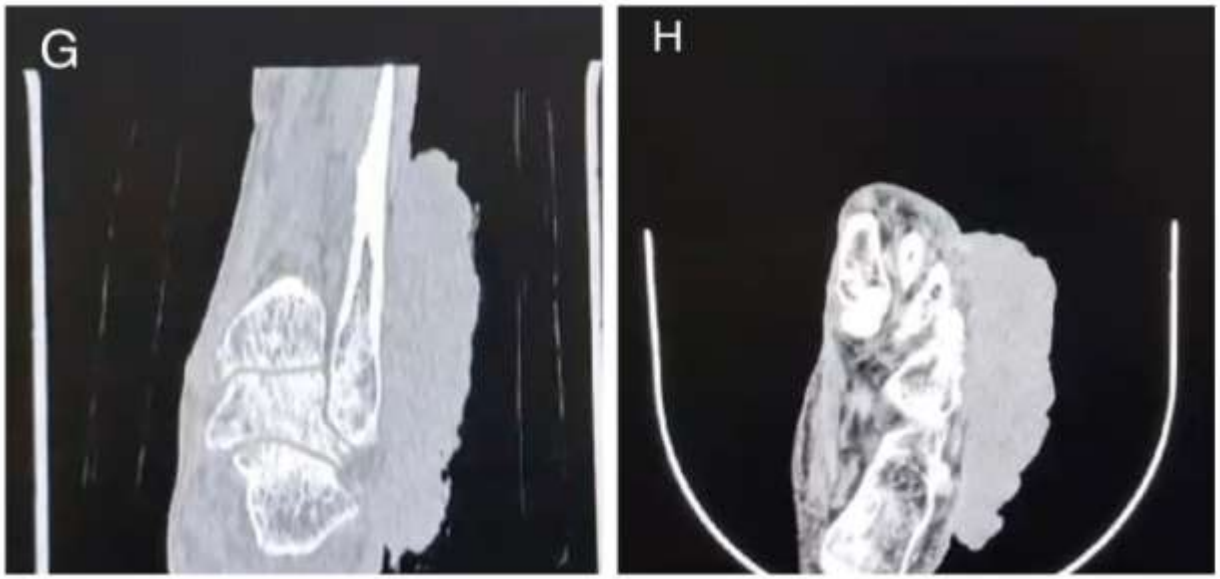

Figure G:-CT LEFT ANKLE WITH FOOT showing heterogenous lobulated soft tissue density mass (Coronal view).
Figure H:- CT LEFT Ankle With Foot showing heterogenous soft tissue denity mass (axial view).

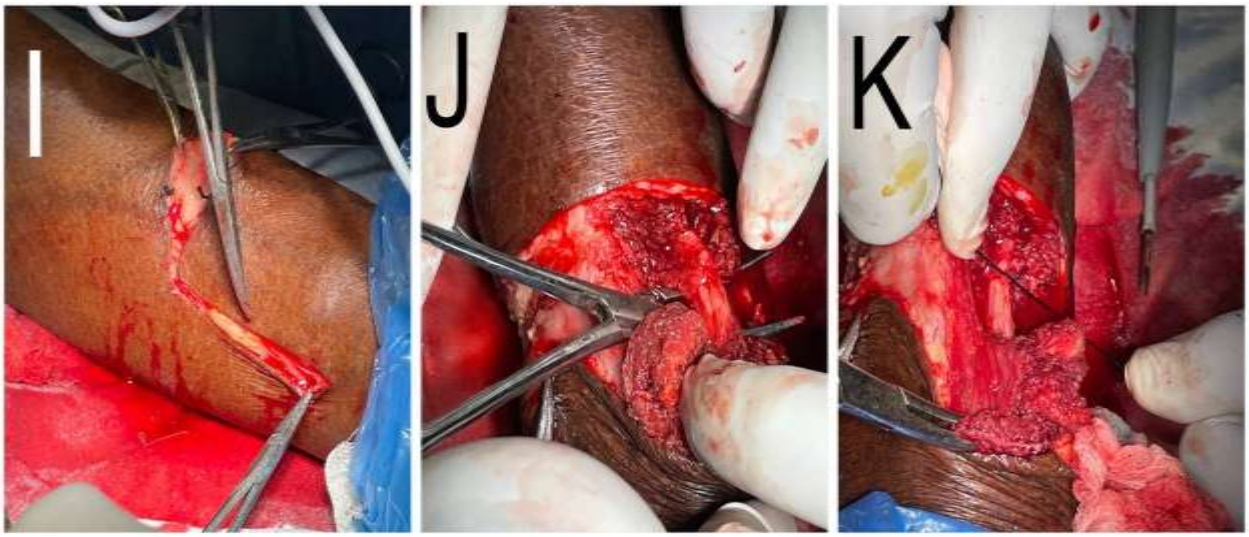

Figure I:-Skin Incision -12cm from tibial tuberosity and making of posterior flap(1.5 times than lenth of transverse insicion)
Figure J,K:- Identification of anterior pedicle and ligation of anterior pedicle.

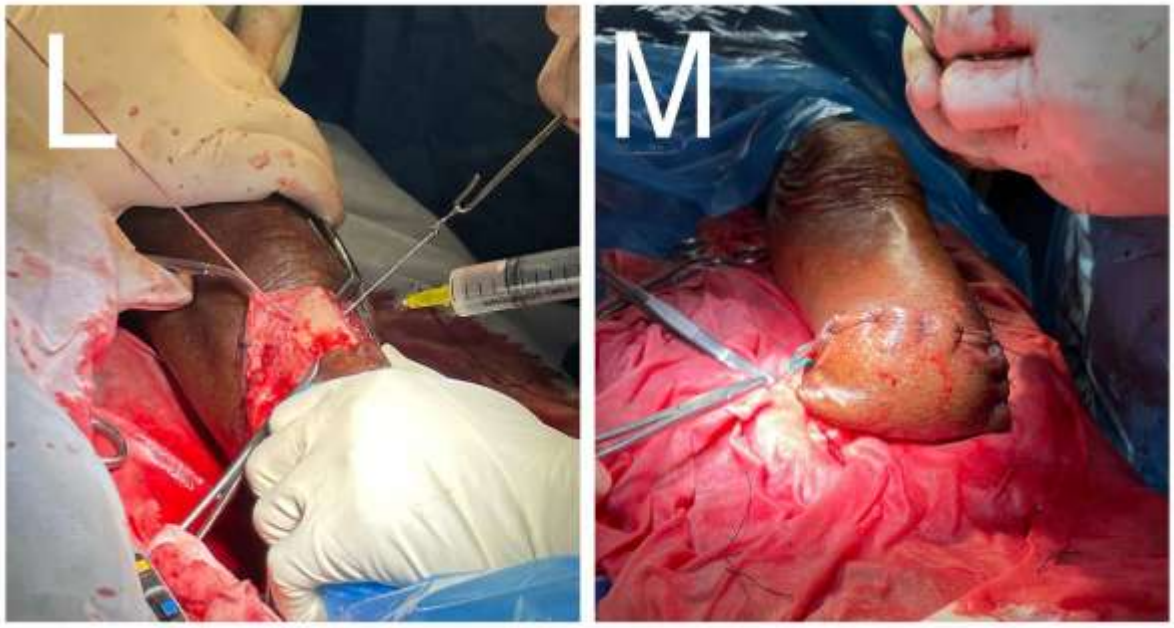

Figure L:- Cutting of TIBIA with gigli saw.
Figure M:-Placement of CRD drain and closure of posterior flap.