Original Research Paper Volume - 4 | Issue - 1 | January - 2020 | ISSN (O) : 2663-0729 | ISSN (P) : 2663-0710

A CASE OF RECURRENT PLEOMORPHIC UNDIFFERENTIATED SARCOMA OF THE RIGHT ELBOW

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ABSTRACT

PLEOMORPHIC UNDIFFERENTIATED SARCOMA PREVIOUSLY KNOWN AS MALIGNANT FIBROUS HISTIOCYTOMA IS A TYPE OF SOFT TISSUE SARCOMA. WE REPORT A CASE OF 76 YEAR OLD MAN WHO PRESENTED WITH A RECURRENT SWELLING IN THE RIGHT ELBOW FOR 6 MONTHS. INITIALLY DONE CORE NEEDLE BIOPSY AND WIDE LOCAL EXCISION SHOWED PLEMORPHIC UNDIFFERENTIATED SARCOMA. PATIENT HAD RECURRENCE AND HENCE WIDE LOCAL EXCISION AND SPLIT SKIN GRAFTING WAS PERFORMED FOR THE PATIENT.

KEYWORD

PLEOMORPHIC, HISTIOCYTOMA, EXCISION, RADIOTHERAPY

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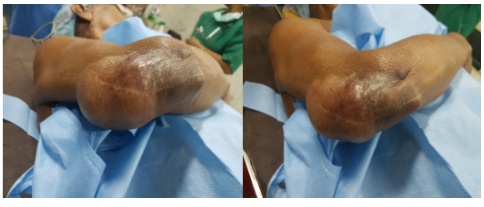
INTRODUCTION:

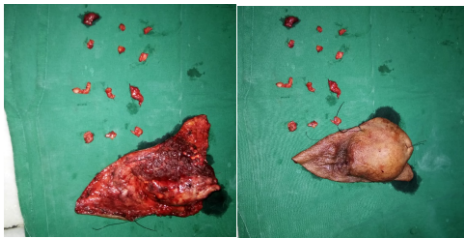
PLEOMORPHIC UNDIFFERENTIATED SARCOMA PREVIOUSLY KNOWN AS MALIGNANT FIBROUS HISTIOCYTOMA IS A TYPE OF SOFT TISSUE SARCOMA. IT IS THE SECOND MOST COMMON SOFT TISSUE SARCOMA IN ADULTS, WITH AN INCIDENCE OF 0.88 CASES PER 1,00,000 ANNUALLY AND INCIDENCE INCREASES WITH AGE. IT AFFECTS BOTH MEN AND WOMEN BUT MORE COMMONLY IN MEN (2:1, MALE:FEMALE). MOST CASES OCCUR IN THE DEEP SOFT TISSUES OF THE EXTREMITIES AND TRUNK. METASTATIC DISEASE IS IDENTIFIED IN 5% OF PATIENTS AT PRESENTATION.

CASE REPORT:

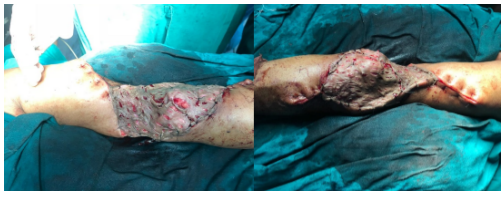
A 76 YEAR OLD MALE CAME WITH COMPLAINTS OF SWELLING IN THE RIGHT ELBOW FOR 6 MONTHS WHICH WAS ASSOCIATED WITH PAIN. PATIENT HAD NO KNOWN CO MORBIDITIES. ON CLINICAL EXAMINATION IT WAS A MULTILOBULATED SWELLING IN THE RIGHT POSTERIOR ASPECT OF THE ELBOW. SKIN APPEARED TO BE FREE OF THE SWELLING. THERE WAS NO DISTAL NEUROVASCULAR DEFICIT. CORE NEEDLE BIOPSY WAS PERFORMED WHICH SHOWED MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR WITH RHABDOID DIFFERENTIATION. WIDE LOCAL EXCSION WAS DONE AND THE TISSUE SENT FOR HISTOPATHOLOGICAL EXAMINATION SHOWED PLEOMORPHIC UNDIFFERENTIATED SARCOMA WHICH WAS S100 AND CK NEGATIVE. AFTER 6 MONTHS PATIENT HAD RECURRENT SWELLING IN THE SAME REGION (1). MRI OF RIGHT ELBOW WAS DONE WHICH SHOWED A LARGE HETEROGENOUS SOFT TISSUE INTENSITY MASS LESION OF SIZE 9 X 5 X 5 CM POSSIBLY COMPRESSING THE ADJACENT ULNAR NERVE. HRCT CHEST AND USG ABDOMEN WERE DONE AND POSSIBILITY OF METASTASIS WAS RULED OUT. PATIENT AGAIN UNDERWENT WIDE LOCAL EXCISION (2) WITH SPLIT SKIN GRAFT (3). PATIENT WAS DISCHARGED WITH SPLIT SKIN GRAFT.



(1) PRE OP PHOTOS OF THE SWELLING.



(2) INTRA OP PICTURES OF THE SWELLING



(3) POST OPERATIVE PICTURES FOLLOWING WIDE LOCAL EXCISION AND SPLIT SKIN GRAFT

DISSCUSION:

UNDIFFERENTIATED PLEOMORPHIC SARCOMA IS A TYPE OF SOFT TISSUE SARCOMA PREVIOUSLY KNOWN AS MALIGNANT FIBROUS HISTIOCYTOMA. SOFT TISSUE TUMOURS FOR WHICH LINE OF DIFFERENTIATION IS DEBATABLE IS CLASSIFIED UNDER PLEOMORPHIC

UNDIFFERENTIATED SARCOMA. RECLASSIFICATION OF MANY TUMOURS IN THIS GROUP AFFORDED BETTER PROGNOSTICATION BUT TRADITIONAL TREATMENTS STILL APPLY.

UNDIFFERENTIATED PLEOMORPHIC SARCOMA ARE FURTHER SUBDIVIDED INTO 5 TYPES: (1) STORIFORM-PLEOMORPHIC, (2) MYXOID (MYXOFIBROSARCOMA), (3) GIANT CELL (MALIGNANT GIANT CELL TUMOR OF SOFT PARTS), (4) INFLAMMATORY, AND (5) ANGIOMATOID.

IT IS THE SECOND MOST COMMON SOFT TISSUE SARCOMA IN ADULTS, WITH AN INCIDENCE OF 0.88 CASES PER 1,00,000 ANNUALLY AND INCIDENCE INCREASES WITH AGE. IT AFFECTS BOTH MEN AND WOMEN. MOST CASES OCCUR IN THE DEEP SOFT TISSUES OF THE EXTREMITIES AND TRUNK. METASTATIC DISEASE IS IDENTIFIED IN 5% OF PATIENTS AT PRESENTATION. MYXOFIBROSARCOMA IS THE MOST COMMON SARCOMA OF OLDER ADULTS, WITH A PEAK INCIDENCE IN THE 7TH AND 8TH DECADES OF LIFE AND A SLIGHT MALE PREDOMINANCE. MYXOFIBROSARCOMA ALMOST ALWAYS ARISES IN SUBCUTANEOUS AND DEEP SOFT TISSUES OF THE EXTREMITIES, AFFECTING LOWER LIMBS AND LIMB GIRDLES MORE OFTEN THAN THE UPPER LIMBS. TRUE CASES OF UNDIFFERENTIATED PLEOMORPHIC SARCOMA WITH GIANT CELLS AND UNDIFFERENTIATED PLEOMORPHIC SARCOMA WITH PROMINENT INFLAMMATION ARE EXCEEDINGLY RARE. THEY ARISE IN OLDER INDIVIDUALS AND AFFECT BOTH GENDERS EQUALLY. THE FORMER APPEARS TO MAINLY AFFECT DISTAL EXTREMITIES AND TRUNK, WHILE THE LATTER OCCURS MOST OFTEN IN THE RETROPERITONEUM OR ABDOMEN. THE BEHAVIOUR OF PLEOMORPHIC UNDIFFERENTIATED SARCOMA IS VERY AGGRESSIVE AND PROGNOSIS IS POOR.

THE MAINSTAY TREATMENT FOR ALL SOFT TISSUE SARCOMAS INCLUDING UNDIFFERENTIATED PLEOMORPHIC SARCOMA IS WIDE LOCAL EXCISION WITH 2CM MARGINS OF UNINVOLVED TISSUE. THE MOST RADICAL TREATMENT FOR PLEOMORPHIC SARCOMAS OF THE EXTREMITY IS AMPUTATION, BUT RATES OF OVERALL SURVIVAL ARE NO BETTER AFTER AMPUTATION THAN AFTER LIMB-SPARING THERAPY. ADJUVANT RADIOTHERAPY IS OFFERED FOR SOFT TISSUE SARCOMA, INCLUDING PLEOMORPHIC SARCOMAS, IN WHICH RESECTION WITH WIDELY NEGATIVE MARGINS IS NOT POSSIBLE. CHEMOTHERAPY IS GENERALLY RESERVED FOR METASTATIC DISEASE. REGIONAL LYMPH NODE METASTASES FROM PLEOMORPHIC SARCOMAS ARE RARE. THE MOST COMMON SITE FOR METASTASES FROM PLEOMORPHIC SARCOMAS IS THE LUNG, AND ALSO BONE AND LIVER. RECURRENCE RATES ARE HIGH WHICH RANGE FROM 25% TO 75%.

CONCLUSION:

UNDIFFERENTIATED PLEOMORPHIC SARCOMA IS A AGGRESSIVE TYPE OF SOFT TISSUE SARCOMA. THE TREATMENT OF UNDIFFERENTIATED PLEOMORPHIC SARCOMA REMAINS THE SAME AS OTHER SOFT TISSUE SARCOMAS. THE PROGNOSIS IS POOR BECAUSE IT IS AN AGGRESSIVE TUMOUR WITH HIGH RECURRENCE RATES. THE ABOVE REPORTED PATIENT IS A CASE OF RECURRENCE WHO UNDER WENT WIDE LOCAL EXCISION WITH SPLIT SKIN GRAFT AND WAS DISCHARGED.

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