

ZEBRA BODIES ON ELECTRON MICROSCOPY; A CASE REPORT

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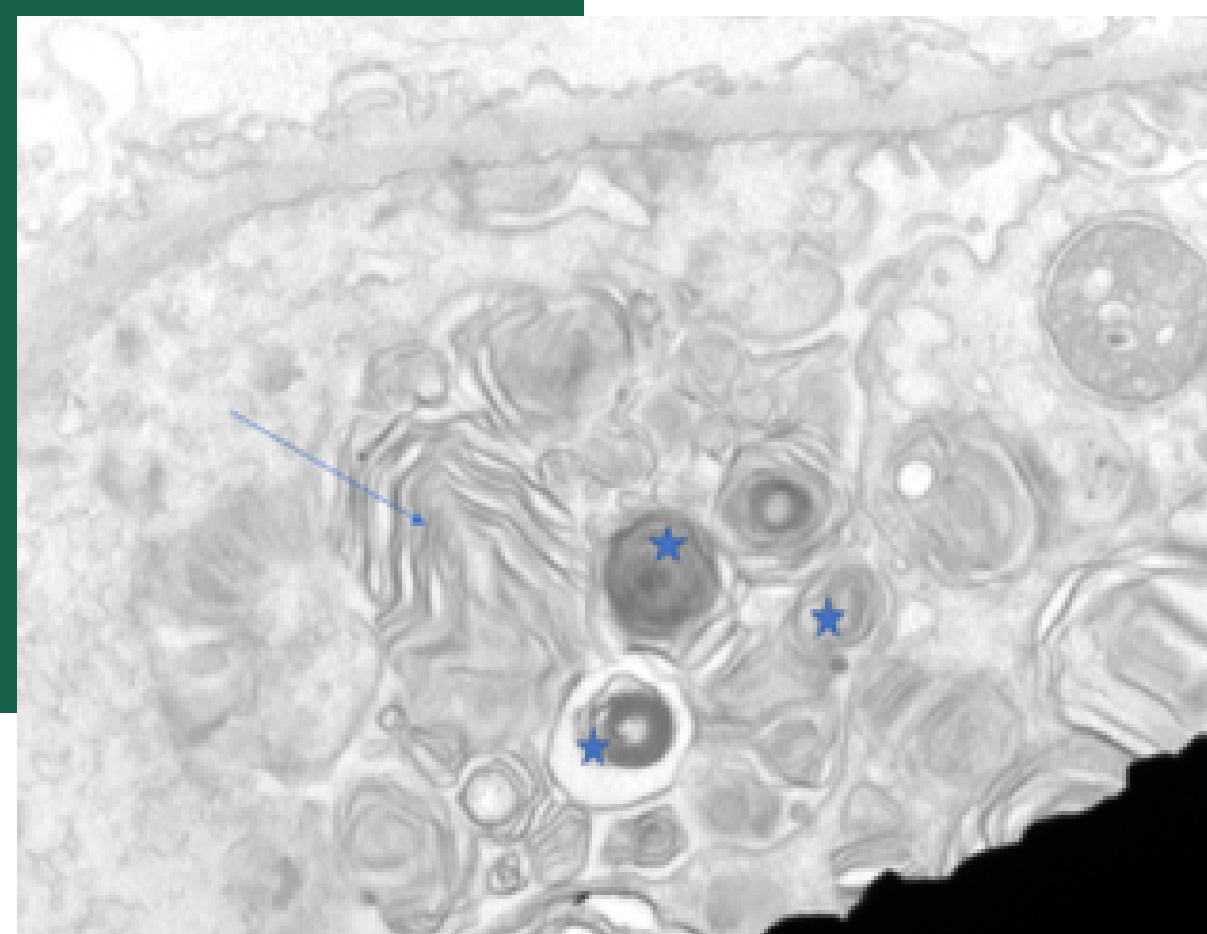
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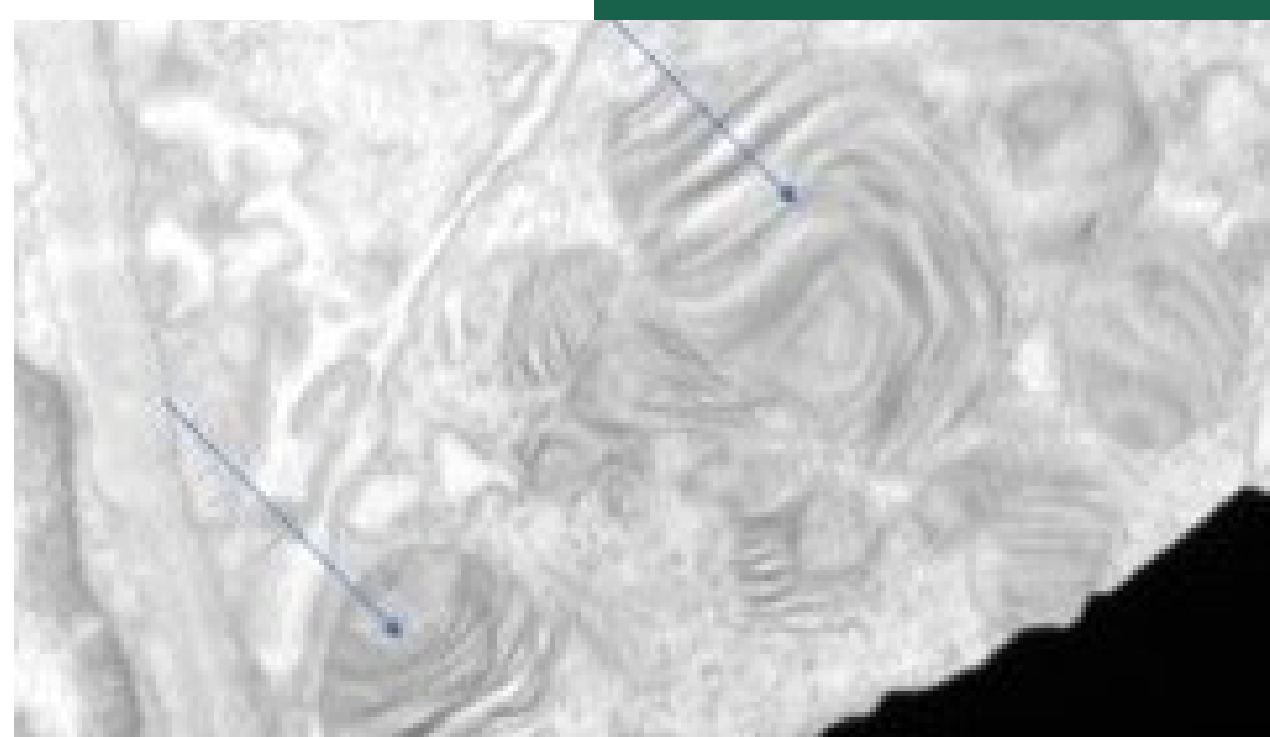
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MYELIN FIGURES (ASTERISK) ON ELECTRON MICROSCOPY



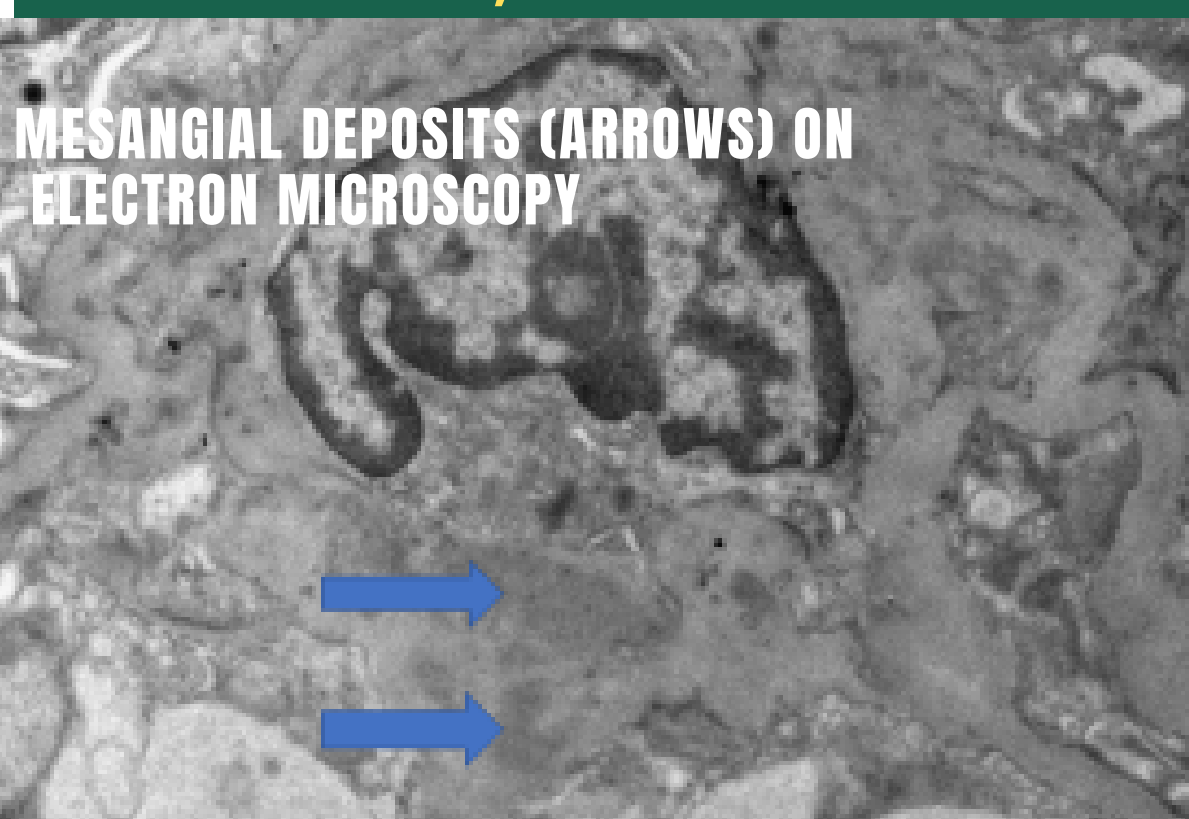
ZEBRA BODIES (ARROWS) ON ELECTRON MICROSCOPY

INTRODUCTION: KIDNEY BIOPSY IS A VALUABLE AND OFTEN INDISPENSABLE TOOL GUIDING THE DIAGNOSIS AND OPTIMAL APPROACH TO THE TREATMENT OF INTRINSIC KIDNEY DISEASES.

CASE REPORT: A 61-YEAR OLD MAN WITH SERONEGATIVE RHEUMATOID ARTHRITIS, FREQUENT ATTACKS OF URIC ARTHRITIS AND A FOUR-YEAR DIABETES PRESENTED WITH ACUTE NEPHRITIC SYNDROME. KIDNEY BIOPSY REVEALED IGA NEPHROPATHY OF MINIMAL ACTIVITY AND NO CHRONICITY DESCRIBED BY OXFORD SCORE M0 E0 S0 TO C0, AND MINIMAL ACUTE TUBULAR INJURY AND FOCAL INTERSTITIAL NEPHRITIS. ELECTRON MICROSCOPY FOUND VERY SCARCE IGA DEPOSITS THAT WERE PROBABLY NON-PATHOGENETIC AND MYELOID OR ZEBRA BODIES IN THE CYTOPLASM OF PODOCYTES, PRIMARILY SUGGESTIVE FOR FABRY DISEASE.

DISCUSSION: MYELOID OR ZEBRA BODIES ARE A HALLMARK OF GLYCOLIPID STORAGE DISORDERS. DEPOSITS OF GLOBOTRIAOSYL CERAMIDE (GB3) AS LAMELLATED MEMBRANE STRUCTURES WITH AN ONION SKIN APPEARANCE MAY BE FOUND IN ALL GLOMERULAR CELL TYPES, MOST PROMINENTLY PODOCYTES, ARTERIOLAR SMOOTH MUSCLE CELLS, AND TUBULES. HOWEVER, LAMELLAR INCLUSIONS HAVE BEEN DESCRIBED IN OTHER CONDITIONS, SUCH AS SILICOSIS, AND WITH THE USE OF CATIONIC AMPHIPHILIC DRUGS SUCH AS ANTIMALARIALS CHLOROQUINE AND HYDROXYCHLOROCHINE, AMPHETAMINE CHLORPHENTERMINE, ANTIHISTAMINE CHLORCYCLIZINE, TRICYCLIC ANTIDEPRESSANTS IMIPRAMINE AND CLOMIPRAMINE, AND ANTIBIOTIC GENTAMYCIN. THE LOCATION OF THE INCLUSIONS IS SOMETIMES HELPFUL IN MAKING THE DISTINCTION BETWEEN THESE DISEASES. LAMELLAR INCLUSIONS ASSOCIATED WITH GENTAMYCIN OCCUR IN PROXIMAL TUBULES, WHEREAS, IN FABRY DISEASE, THE INCLUSIONS ARE MOST STRIKING IN PODOCYTES AND DISTAL TUBULES.

MESANGIAL DEPOSITS (ARROWS) ON ELECTRON MICROSCOPY



CONCLUSION: LEUCOCYTE ALPHA-GALACTOSIDASE A (ALPHA-GAL A) ACTIVITY ENZYMIC ASSAY RULED OUT FABRY DISEASE. SINCE OUR PATIENT HAD BEEN TREATED WITH HYDROXYCHLOROQUINE FOR OVER A DECADE, THE DRUG WAS SUSPENDED. THE PATIENT SIGNIFICANTLY RECOVERED HIS KIDNEY FUNCTION IN THE FOLLOW-UP.