IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861. Volume 11, Issue 4 (Nov.- Dec. 2013), PP 79-82
www.iosrjournals.org

Rheumatoid Arthritis Complicated With Systemic A5myloidosis Diagnosed At Autopsy: A Case Report.

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I. Introduction:

Rheumatoid arthritis (RA) is not commonly reported in Nigeria, a cross sectional survey of 520 inhabitants in a rural setting in the south east of the country found the disease in approximately 17.7% [1], a recent review of 200 consecutive RA patients diagnosed in a rheumatology clinic over a seven year period, concluded that it is not a rare disease in Nigeria [2].The aetiology RA is still unclear, accumulating evidences are clarifying the involvement of genetic and environmental factors in the disease, or the pathogenic mechanism of perpetuating proliferative synovitis and bone destruction[3].Presenting primarily as articular and bone disease, it is the extra articular presentation and complcations however that are responsible for most fatalities [4,5,6]

The most frequent extra articular complications leading to death are overwhelming sepsis, systemic vasculitis and secondary amylodosis [4,5,6] .A review of 234 autopsies carried out in a rheumatology centre in Hungary found that complications were clinically recognised in only 46.6% of the cases[7]. In terms of organ complication, renal complication following vasculitis,amyloidosis, glomerulopathies is common , equally important is heart disease due to amyloidosis and vasculitis; but virtually every organ system may be involved. Secondary amyloidosis is a life-threatening complication of rheumatoid arthritis RA. When amyloidosis sets in, cardiac and renal involvements are poor prognostic signs [8].

II. Case Report:

This is a case report of a 34 year old male relative of a medical doctor who developed progressively worsening polyarthritis starting from the metatarsal joints, it spreads through the ankle joints, knee joints, temporo-mandibular joints and sternoclavicular joints.The pain responded to non steroidal anti inflammatory drugs. Against the advice of his doctor relative, he sought Medicare with traditional healers who inflicted scarification marks above the inflamed joints. By the time the relation intervened about a year after, his condition had grown worse ,the pain was crippling, he had developed macroglossia for which an Ear Nose and throat (ENT) surgeon was consulted but the cause was not found. There was no history of oliguria or anasarca to suggest failure of his kidneys and there was no indication of overwhelming infection.

Investigations:

Terminally, he developed normochromic anemia,his electrolytes urea and creatinine were within normal limits, he had moderate proteinuria but no glycosuria.He did not have a septic workup, rheumatoid factor was not assayed for, neither was his Human Leucocytes Antigen status (HLA) determined. Radiological examinations were not carried out. The patient died suddenly one day and autopsy was requested for.

Autopsy Findings:

General physical examination revealed a rather well nourished young man, mildly pale with an anterior neck mass 4X3X2.5 cm. The cut surface is solid and rather meaty, subcutaneous nodules were not found. The rest of the physical findings were unimpressive.

The parietal pleura on both sides was fibrotic and adhered to the chest wall, the rest of the serous cavities were unimpressive.

There was mild cardiomegaly, the heart weighed 400g and had a rubbery consistency .There was marked left ventricular hypertrophy (2cm), narrowing of the ventricular cavity was appreciated and both papillary muscles and trabecular carnae were rigid. The coronary arteries were patent. Irregular amyloid deposits were seen separating cardiac myocytes with areas of progressive myocytolysis.

Both kidneys displayed mild increment in weight and appear rubbery in consistency when cut open.Microscopically amyloid was seen on the glomerular turfs and around the tubules.

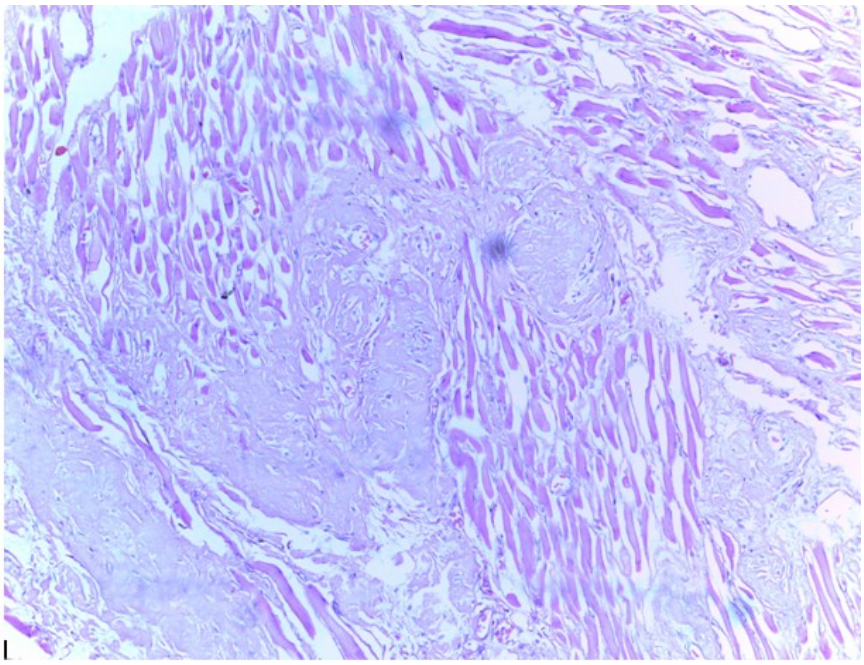
There was macroglossia ,the tongue weighed 350 g ,was firm to rubbery in consistency and microscopically, amyloid tumoral masses were seen deposited between muscle fibres with extensive muscle fibre atrophy .

The thyroid gland was solid rubbery and amyloid was demonstrated around blood vessels.

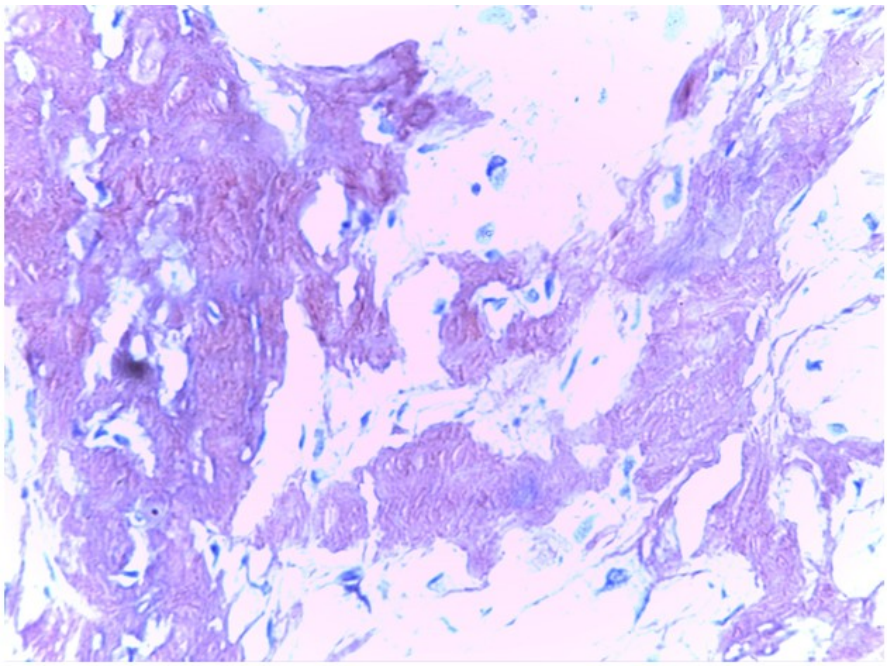
A biopsy of the sternoclavicular joint shows thick inflammatory fibrous pannus, and narrowing of the joint space.

The spleen was not enlarged (160g), none of the of the specific patterns of amyloid deposition was noticed grossly, but microscopically, amyloid was deposited around splenic sinusoid and connective tissue framework.

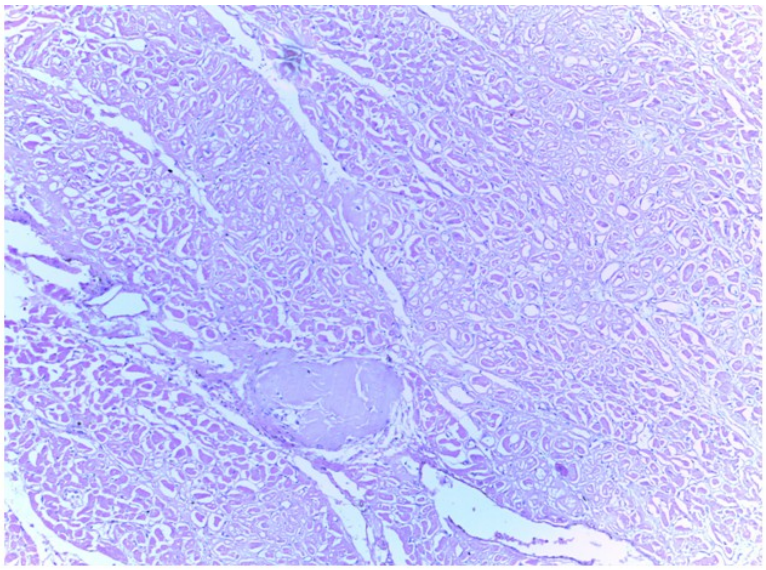
It was concluded that the patient died of secondary amyloidosis, complicating rheumatoid arthritis. The actual cause of death most likely due to cardiac complications as there was no indication of severe renal compromise or evidence of systemic infection or vasculitis which are the commonly reported complications.



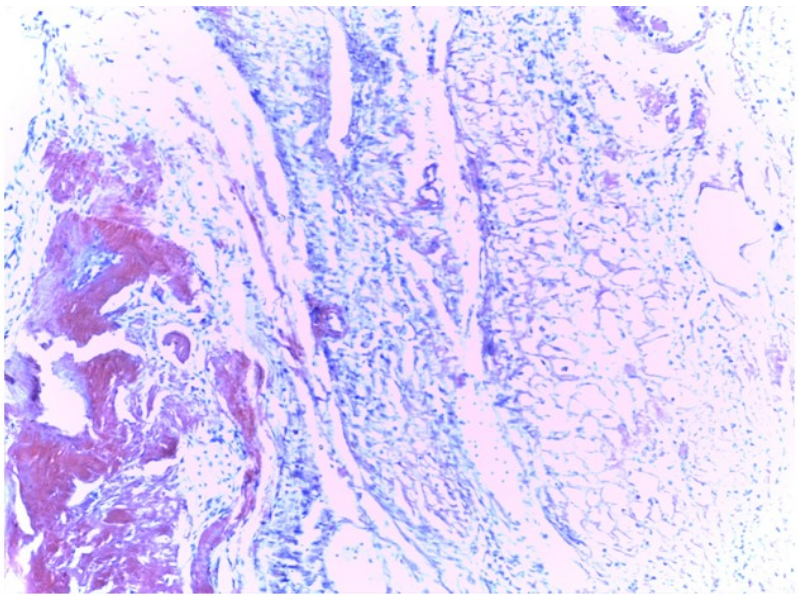
H & E TONGUE x 10 (Amyloid nodules with skeletal muscle atrophy)



Congo red Tongue X 40 (zone of severe amyloid deposit and extensive muscle fiber atrophy)



H& E Cardiac muscle X 10 (amyloid nodule myocytolysis is extensive)



Congo red Heart muscle X 10 showing amyloid deposit

III. Discussion:

A earlier study in an Ibadan cohort presenting with poly arthritis concluded that RA may not be common among Nigerians[9], this seem to have been contradicted by a recent Lagos study[2].Both Ibadan study and the Lagos study employed American rheumatology association ARA criteria for diagnosis of RA. In particular the Ibadan study noted in details the peculiar differences between the Ibadan RA patients and the typical western patients; notable in their patients rheumatic factor RF positivity was low compared to western patients, subcutaneous nodules were fewer, bone and joints changes were less severe. Their conclusion was that chronic antigenemia and immunological challenge occasioned by endemic malaria and other infectious diseases experienced by Ibadan RF patient, may have modified the disease presentation in this environment .Our index patient did not have subcutaneous nodules and had taken to alternative care .Who knows how many RA patients in this environment routinely shun orthodox care for alternative care, because of this preference, the true incidence of this disease in this senvironment is not known. The assertion that the disease is not common may be spurious and may be a reporting bias.

According to Bèly M and colleague who reviewed 161 autopsies carried out in a rheumatology centre between 1970-1999,only 25% of the severe complications in RA were recognised clinically[6] .In a wider study, the same researchers reviewed 234 deaths from rheumatoid arthritis out of 10,860 overall deaths in the same rheumatology centre and found out that only 46.6% of the complications were recognised clinically, however

two thirds of the lethal complications were recognised [7]. It is therefore not surprising that in our index patient, amyloidosis was not recognised as the cause of his macroglossia clinically.

For clinical case detection of secondary amyloidosis in RA to be quick, the clinician must entertain a high index of suspicion and know the limitations and hence choose appropriate diagnostic method. In one reported RA series 91% of the renal biopsies were positive while only 52% of abdominal fat aspirations were positive[10]. Among the commonly employed diagnostic methods are rectal biopsy, renal biopsy and more easily abdominal fat aspiration.

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