

**THE RESPIRATORY SYSTEM**

*Sub*-system: patholoy

Lecture Title: sarcoidosis (restrictive lung Dz )

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***Sarcoidosis # (اللحمانية)***

**Modified**

**Sheet note after # , slides in boxes**

**Remember: Restrictive lung Dz > chronic diffuse interstitial lung Dz > granulomatous > sarcoidsis .**

***What is the sarcoidosis ?***  **\*\*a multisystem disease , unknown etiology , noncaseating granulomatous inflammation in many tissues and organs # any organ in the body .**

 **\*\*bilateral hilar lymphadenopathy or lung involvement (or both), visible on chest radiographs, is the major finding at presentation in most cases .**

**\*\*Eye and skin involvement each occurs in about 25% of cases, and either may occasionally be the presenting feature of the disease.**

***Microscopic findings in granuloma :***

**Schaumann and asteroid bodies inside the granulomas .**

**# schaumann (concentric calcifications , purple in color ) # asteroid (needle-shape structure crystals ) .**

 ***\*\* Extra note : in pathology, Schaumann bodies are calcium and protein inclusions inside of Langhans giant cells as part of a granuloma .***

 ***epidemiology :***

**#no differences between genders** .

***Pathogenesis :***

**#even we said that the causes are unknown .**

**But , Several immunologic abnormalities in sarcoidosis suggest the development of a cell-mediated response to an unidentified antigen. The process is driven by CD4+ helper T cells**

**# in general , helper T cell activate macrophages that make the granuloma ….. , so keep in mind that there is some immunologic abnormalities that suggest development cell mediated response to unidentified antigen by T cells that activate the granulomatous inflammation at the end . (تم استجابة زائدة في الجسم )**

***organs involved :***

***lungs :***

 **lungs…90% of cases # remember the most common manifestation .**

**…with tendency to localize in the connective tissue around bronchioles and pulmonary venules and in the pleura (“lymphangitic” distribution)**

 **# it's similar to the anatomy of lymphatics .**

**…5% to 15% of patients, the granulomas eventually are replaced by diffuse interstitial fibrosis, resulting in a so-called “honeycomb lung”**

**# 5-15% (few) from patients that have lung involvement (5-15 % from 90%) that may progresses to the end-stage lung Dz (honeycomb lung ) , it's extensive عنيف fibrosis , sever form .**

***Lymph nodes :***

**Hilar and paratracheal lymph nodes are enlarged in 75% to 90% of patients, while one-third present with peripheral lymphadenopathy**

**#the second most manifestation .**

***Skin :***

**especially erythema nodosum…sarcoidal granulomas uncommon in these lesions… lupus pernio .**

**#the skin manifestation :**

**1) EN : the most common skin lesion , even though there is little EN in non-caeseating granuloma , it associated with : sarcoidosis , pancolitis , …etc.**

**extra : remember : EN is skin inflammation is located in fatty layer of skin associated with more than one Dz .**

**2) sarcoidal granuloma .**

**3)lupus pernio : important for clinical …..**

**Student asked the doctor : Is this same as systemic lupus erythematosus ?**

**Ans : no , this lupus is different from lupus erythematosus .**

**And keep in mind that we have lupus vulgaris which is TB in skin .**

**These terms will be helpful for us in clinical , the prof said .**



***Eyes and glands :***

**Eye & lacrimal glands…1/5 to 1/2 # nearly 25% of patients (iritis or iridocyclitis, unilateral or bilateral) , also choroiditis, retinitis, optic nerve involvement \* suppression of lacrimation>>>>>sicca syndrome.**

**#the sarcoidosis can affect any part of eye .**

**# sicca syndrome : clinical syndrome that inhibit the lacrimation (xeroophthalmia :جفاف العين ) and salivation (xerostomia : جفاف الشفاه ) .
Also , it can occur in Sjogren's syndrome ( autoimmune Dz affect salivary glands ) .**

 **Unilateral or bilateral parotitis with painful enlargement #swelling in <10%...xerostomia**

**\*\*Combined uveoparotid involvement is designated Mikulicz syndrome**

 **# uvea in eye and parotid gland ( salivary gland ) we call it mikulicz syndrome #we said that it can affect any part of eye (uvea )**

 **Extra note : uvea is the pigmented layer of the eye, lying beneath the sclera and cornea, and comprising the iris, choroid, and ciliary body.**

***Others :***

**Spleen, liver and bone marrow: commonly involved by microscopic granulomas but less commonly with clinical manifestations**

**# microscopic more than clinical ??? that u will find structural changes under microscope more than symptoms such as hepatitis for ex** .

***Hypercalcemia…Why?***

**# if we back to the immunological abnormalities that lead to sarcoidosis , we have to know that the activation of macrophages can produce the active form of vit.D which will increase the calcium in Bld , and this hypercalcemia can lead to metastatic calcification or dystrophic calcification .**

 **#NOTE : is not necessary for this condition(sarcoidosis ) to occur in lung even though is the most common manifestation , it may occur only in skin or only in eye .**

**(بمعنى انها ممكن تكون بالعين مثلا بدون الرئة وهكذا )**

 ***clinical notes :***

**\*\*Many are asymptomatic…incidentally on x-ray or autopsy: bilateral hilar lymphadenopathy**

**#it can appear on x-ray in any organ , bcz it affects any organ .**

**\*\* Any organ of the previously mentioned may be the presenting one**

**\*\* In 2/3s of symptomatic patients: respiratory (dyspnea, dry cough) or constitutional signs & symptoms (fever, weight loss, night sweats etc.)**

**#the prof said that we must be familiar to night sweats causes "some of them " :**

1. **hodgkin's lymphoma**
2. **TB**
3. **Sarcoidosis**
4. **Brucellosis (malta fever ) .**

***\*\* For diagnosis:***

**1- Clinical, radiographic and biopsy findings after excluding other causes especially TB may cause noncaseating granulomas.**

**# first in diagnosis , u have to rule out everything that may involve or any known cause for granulomatous inflammation (caseating or non-caseating ) such as : TB (caseating granuloma) , fungal infection hypersensitivity pneumonitis (non-caseating) , occupational lung Dz (Berylliosis or chronic beryllium Dz ).**

 **2-ACE is increased in the blood and in bronchoalveolar lavage…about 60% of cases**

**3-CD4+ T cells are increased around the granulomas and in bronchoalveolar lavage while there is peripheral T cytopenia**

**# what is the bronchoalveolar lavage ?**

**Broncoscopy (تنظير للقصبات )**

**is a**[**medical procedure**](https://en.wikipedia.org/wiki/Medical_procedure)**in which it involves fluid is squirted into a small part of the lung and then collected for examination (cytopathology >> have cells and fluids for diagnosis ). It is typically performed to**[**diagnose**](https://en.wikipedia.org/wiki/Medical_diagnosis)[**lung disease**](https://en.wikipedia.org/wiki/Lung_disease)**.**

**# ACE high in Bld and bronchoalveolar lavage but CD4+ T cells high in Bld and few in bronchoalveolar lavage .**



 **#the course is different from patient to patient .**

#معظم ا**لحالات بصيرremission .**

**# ممكن يرجع ينصاب فيها المريض وممكن لأ**

**#cor pulmoale : lung fibrosis .**

What is the Lofgren syndrome ?

**# It is acute presentation of :**

1. **hilar lymphadenopathy**
2. **erythema nodosum**
3. **arthralgia or arthritis**

**best luck for all .**