Journal Homepage: -www.journalijar.com
INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)
Article DOI:10.21474/IJAR01/8457
DOI URL: http://dx.doi.org/10.21474/IJAR01/8457
ISSN NO. 2320-5407

RESEARCH ARTICLE

GOOD'S SYNDROME:REPORT OF CASE.

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Manuscript Info

Manuscript History
Received: 02 December 2018
Final Accepted: 04 January 2019
Published: February 2019

Key words:-
Hypogammaglobulinemia ; Intravenous gammaglobulin; Thymoma.

Abstract

Good's syndrome is defined by the association of thymoma, hypogammaglobulinemia and recurrent infections especially bronchopulmonary and sinus. We report the case of a 53 years old man , with a history of recurrent respiratory infections, hospitalized in June 2017 for bronchial syndrome dragging with radiographic front chest presence of bilateral alveolar syndrome and latérotrachéale opacity right, supplemented by a thoracic scan, which objectified the chance discovery of a regular meditational tissue mass without locoregional malignancy, with no clinical and electrophysiological signs of myasthenia gravis. The patient was operated for his médiatisnal mass which histological study was in favor of a thymoma type A, the outcome was favorable. In our patient the association between thymoma and hypogammaglobulinemia and bronchopulmonary infection severe and recurrent led to the diagnosis of Good's syndrome. The patient was put under appropriate antibiotic therapy combined with weekly infusions of intravenous immunoglobulin, with a positive trend, achieving significant clinical improvement, biological and radiological. Good's syndrome represents only 5% of parathymiques diseases, but its research is essential because it allows effective treatment of infections, avoiding lung destruction and the use of lung surgery.

Introduction:-

The good's syndrome was described in 1954 by Good and al as a pathological entity thymoma complicated hypogammaglobulinemia. [1] Although the symptoms of myasthenia pure red cell aplasia, which often occur with thymoma, generally improved by thymectomy, good syndrome appears to be progressive, although thymectomy and / or treatment with corticosteroids are applied. [2]

Case report

We report the case of a 53 years old man, no known chronic smoking followed for type 2 diabetes on metformin well balanced, with a history of respiratory infection repeatedly hospitalized in June 2017 for bronchial syndrome with dragging in chest radiograph the presence of bilateral alveolar syndrome and latérotrachéale right opacity (Fig 1), a pneumologique assessment was performed on the objectifying scannographic plane small bronchi expansion homes with some alveolar condensation homes frosted bilateral and discovery of an earlier médiatisnale tissue mass

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without signs of locoregional malignancy (Fig 2). A tumor markers and bHCGalphafoetoprotein was negative. The assay and LDH B2microglobuline came back negative, with absence of clinical signs and electrophysiological myasthenia gravis. The patient was operated for his médiatisnale mass which histological study was in favor of a thymoma Type A classification of the World Health Organization thymoma, the outcome was favorable under antibiotic therapy.

After Three months , the patient was readmitted for an array of severe recurrent infectious pneumonitis operating in a context of fever and impaired general condition, clinical examination revealed a polypnéique patient, tachycardia, with desaturation in the ambient air 85 % febrile at 39 ° C with a weight loss of 15 kg, the pleuropulmonary examination revealed the presence of bilateral rales sounding diffuse. There was no lymphadenopathy or organomegaly palpable.

Chest radiograph objectifying a bilateral diffuse alveolar syndrome , CT scan (Fig 3) showed the presence of a reticulo nodular syndrome affecting both upper lobes left and right and the left lower lobe associated with an infiltrate diffuse and homogeneous frosted glass, with thickening peribronchovascular, associated with mediastinaladenopathies upper pre tracheal infracentimétrique without pleuropéricardique effusion.

A phtisioloqique balance with the search Koch bacillus on direct examination 03 consecutive days in the morning came back negative sputum.

Bronchoscopy objectified an inflammatory aspect of the bronchial mucosa, with completion of purulent secretions from the right lower lobe. Including microbiological study to highlight the presence of Streptococcus pneumoniae sensitive to ceftriaxone and ciprofloxacin.

A biology, there was an inflammatory syndrome with leukocytosis 13.470 g / l, with 80% neutrophils, 10% lymphocytes, 08% monocytes, 01% basophils and 01% eosinophils, a C-reactive protein 96 mg / L and the absence of anemia. Serum electrolytes also objectified hypoprotidemia to 49G / l.

Severe infectious clinical history evoked the presence of immune deficiency, HIV status was negative, serum protein electrophoresis showed hypogammaglobulinemia (1.1g / L) with a drop of immunoglobulinsIgG, IgM and IgA.

The lymphocyte count obtained from circulating blood lymphocytes, showed the absence of B lymphocyte population, an increase in the CD8 T lymphocyte population.

The search for antinuclear antibodies, anti-DNA antibody was negative. Bone marrow was normal.

In our patient, the association betwenthymoma and hypogammaglobulinemia ,bronchopulmonary infection and severe recurrent led to the diagnosis of Good's syndrome.

Under antibiotic therapy (ceftriaxone and ciprofloxacin) associated with weekly infusions of 5 g of intravenous immunoglobulin, evolution was satisfactory, achieving a marked clinical improvement, biological and radiological.

Fig 1:-chest radiograph objectified the presence of bilateral alveolar syndrome and latérotrachéale right opacity

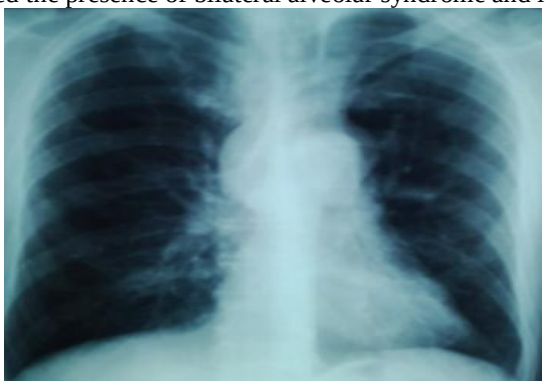


Fig 2:-Chest scan objectified bilateral alveolar condensation and mediational mass

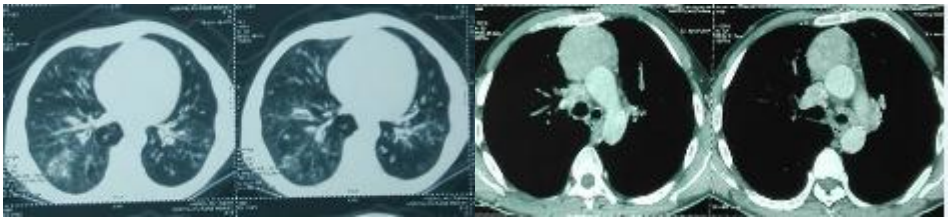
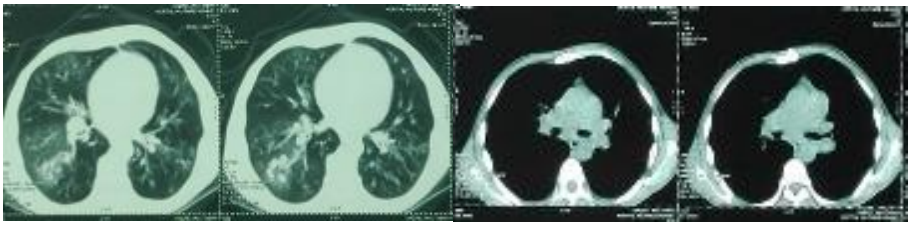


Fig 3:-Chest scan objectified reticulo nodular syndrome affecting both upper lobes left and right and the left lower lobe associated with an infiltrate diffuse and homogeneous frosted glass



Discussion:-

good's syndrome is a rare disease that is responsible for recurrent bacterial, viral or fungal in 95% of cases. The pathophysiology of this syndrome remains unclear, it combines hypogammaglobulinemia, the absence or presence of low levels of B cells in the blood and defects in cell-mediated immunity [3], the syndrome patients have increased susceptibility to sinus and lung infections, pure red cell aplasia is also often associated with particularly good's syndrome in thymoma Type AB.

Our case is a reminder clinical characteristics (thymoma, severe and recurrent infection bronchopulmonary, bronchiectasis) and biological (hypogammaglobulinemia, absence of circulating B lymphocytes) Good syndrome. Bronchiectasis are secondary to severe bronchopulmonary infections, signs of poor immunity.

As in patients with hypogammaglobulinemia, infections are mainly bacterial pyogenic germs. An opportunistic infection Cumpylobacter fetus in septicemic form [3] and mucocutaneous candidiasis [4] however, have been observed.

In our patient, as reported in the literature [5], thymectomy is usually ineffective immune deficiency. The discovery of I'hypogammaglobulinémie almost contemporaneously to the diagnosis of thymoma, has introduced treatment with intravenous immunoglobulin, which has made a major contribution to the healing of the lung.

The association between thymoma and autoimmune disease vector, found in 74% of cases [6], Good syndrome represents only 5% of parathymic syndromes [7]. These are numerous and include myasthenia gravis, PRCA, hemolytic anemia autoimmune, dermatomyositis, systemic lupus and rheumatoid arthritis. [7] An association between a thymoma, myasthenia gravis and polymyositis has also been reported [8].

Thymomas associated with Good's syndrome are mild in 90% of cases and are mostly spindle cell. On the immune plan patients Good syndrome have cutaneous anergy, lack of circulating B lymphocytes and an expansion of the lymphocyte subpopulation CD8 positive T. The inversion of the T4 / T8 ratio is mainly secondary to the expansion of the subpopulation T CD8 positive. Rashal et al. [9] showed that the CD4 and CD8 T lymphocytes express the marker CD45R0, suggesting the predominance of lymphocyte sub-population of T-type memory. The absence of circulating B cells appears to come from a very early blocking differentiation B to the spinal level [3].

Conclusion:-

The discovery of hypogammaglobulinemia in adults involves seeking other conditions especially a syndrome immunodécitaire non-HIV, a lymphoproliferative syndrome with in particular chronic lymphocytic leukemia, non-

Hodgkin's lymphoma or non-secreting myeloma. Diagnosis Good syndrome allowed to offer additional treatment with intravenous immunoglobulin, which would probably have avoided the lung surgery in our patient.

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