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Diagnostic and Therapeutic Approach to HIV-Kaposi's Sarcoma Patients- Case Report

SUMMARY

Background/Aim: *The Acquired Immune Deficiency Syndrome (AIDS) is an immunologic entity, which is due to an RNA virus. AIDS is relatively easy transmitted and has a poor prognosis. The main mechanism of this Syndrome is characterized by the destruction and diminishing of T4 lymphocytes or helper/suppressor T-cells. Kaposi's sarcoma is a neoplasm due to human herpes HHV-8. Kaposi's sarcoma is one of the entities that determines the disease of AIDS. Kaposi's sarcoma appears as skin lesions and attacks the oral cavity and lymph nodes.* ***Case report***: *A male patient, with HIV-syndrome, was referred to the Special Infection Clinic of the University Hospital with a chief complaint of having two lesions, in the oral cavity and in the right ear. The histological report revealed the diagnosis of vascular tumor having morphological and immuno-histochemical characteristics compatible with Kaposi's sarcoma.* ***Conclusions:*** *The doctor must be suspicious when lesions are observed, particularly in the oral cavity. A biopsy must be taken in order to obtain the correct diagnosis and examine the possibility of the presence of Kaposi's sarcoma or AIDS.*

Key words: AIDS, HIV-Infection, Kaposi's Sarcoma

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CASE REPORT (CR)
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Introduction

The Acquired Immune Deficiency Syndrome (AIDS) is an immunologic entity caused by the RNA virus. AIDS is relatively easy transmitted disease and has a poor prognosis. The main mechanism of this syndrome is characterized by destruction and diminishing of T4-lymphocytes or helper/suppressor T-cells[1]. The destruction of these cells leads to the disorganization and general reduction of the immune defense mechanisms of the organism. In the territory of inadequate immune defense, there is a growing number of opportunistic infections and neoplasms, which influence a clinical picture of the disease.

Mobilization of state, scientific, social or other population groups, which are involved in the whole problem, has significantly altered the initial epidemiological data on the HIV-infection. However, the entire prevalence of HIV-infection is not radically eradicated. This is obviously due to the fact that transmission of the HIV virus is done through established habits and archetypes of experiential functions, which constitute a way of life and are difficult to change. The disease is rapidly spreading, the HIV-virus attacks every person with whom it comes into contact, under appropriate conditions, and extends in every geographic area of the planet. World Health Organization Specialists predict that in 2020 HIV-patients will be 40 million and AIDS-patients respectively about 10 million.

The aim of this case report is to introduce therapeutic approach to HIV- Kaposi's sarcoma patients.

Case Report

A male patient, 32 years old with HIV-syndrome, was referred to the Special Infection Clinic of the University Hospital AHEPA in Thessaloniki, with a chief complaint of having two lesions, in the oral cavity and in the right ear. He was anxious and in bad mood due to his state. He stated that he was feeling intense discomfort in the mouth while eating, and he was frightened "that something bad was happening" to him.

Intraoral clinical examination revealed two red-purple nodules, about 5mm in diameter, the larger one situated in the right palate approximately in the first right upper molar region, half way to the midline, and a smaller one of the same color in the left side of the palate, near the midline (Figure 1). A third nodule was also identified behind the right ear, being of a round shape, reddish, and about 25mm in diameter. All three lesions were free of pain on palpation (Figure 2).

Figure 1. Two red-purple nodules half way to the midline

Figure 2. A nodule identified behind the right ear

A biopsy was performed and a tissue specimen was taken from the large lesion of the right palate and was sent for histology to the Department of Pathology, University Clinic of the AHEPA hospital. The histological report revealed the diagnosis of vascular tumor having morphological and immuno-histochemical characteristics compatible with Kaposi sarcoma (Figures 3-5).

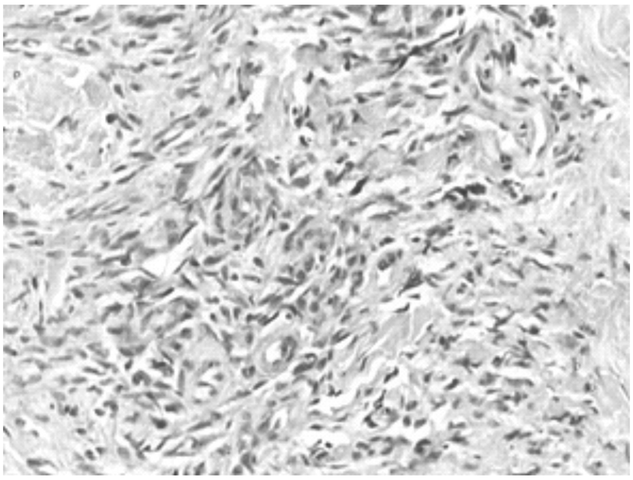

Figure 3. Many small slit-like vessels and spindle cells

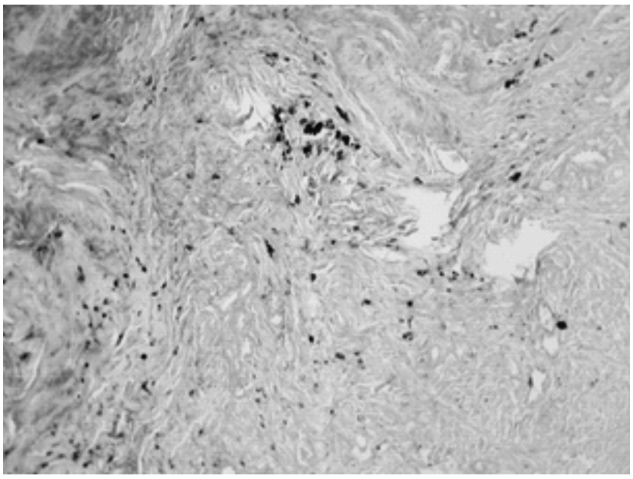

Figure 4. Granular deposits of hemosiderin

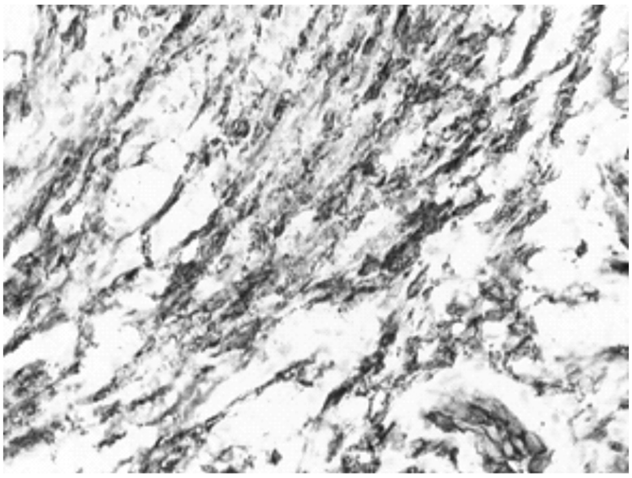

Figure 5. Endothelial and spindle cells - positivity for CD34

According to the results of histological examination, the patient was referred to the Oncology Department for further treatment, according to the protocol of International treatment standards. The patient was referred to the Radiology Department and a CT-Scan imaging of the head was also performed. The results of CT-Scan showed no focal lesions in the cerebral hemispheres and in the cerebellum. Furthermore the CT-Scan confirmed the absence of hematoma or cerebral hemorrhage, and without the points of displacement of the midline structures (Figure 6).

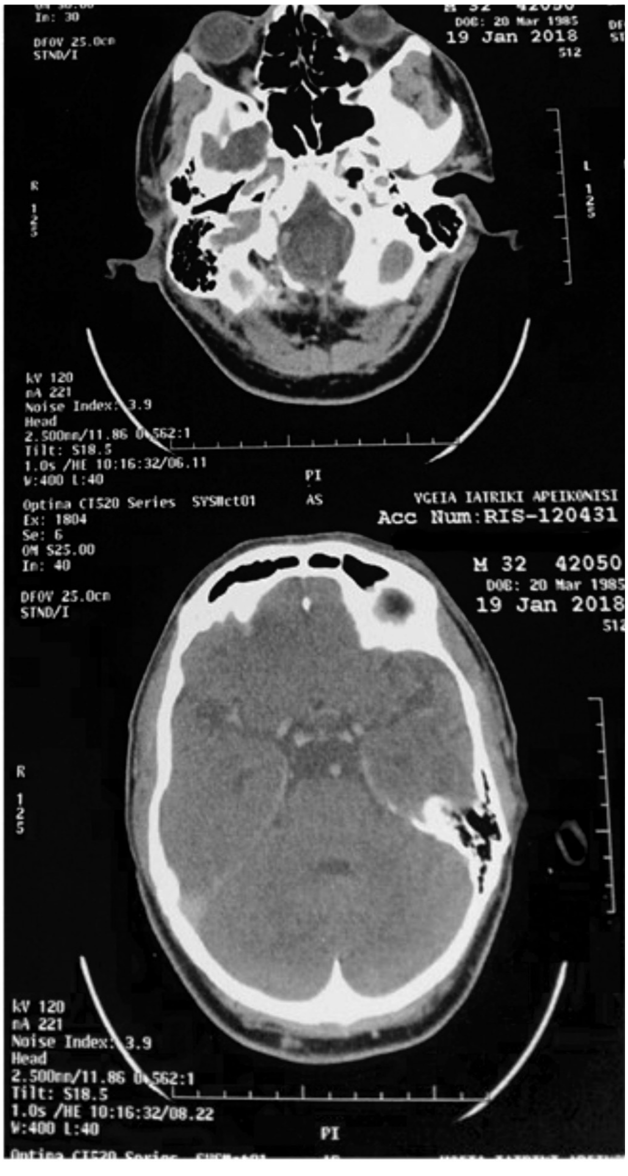

Figure 6. CT-Scan

Chemotherapy was decided by oncologists as a treatment choice. Four cycles have been done so far and two more circles remain. The results so far are encouraging.

Discussion

Hungarian dermatologist Moritz Kaposi, who was working at the University of Vienna, in 1872 first described this lesion[5]. It appears with skin lesions and attacks the oral cavity, lymph nodes and bumps. In 1994 discovery was made that this neoplasm is caused by virus. It has been categorized in four clinical forms: classic form; endemic form; Kaposi's sarcoma associated with suppression of the immune system; and Kaposi's sarcoma associated with AIDS[6].

A clinical picture of the disease is an extremely complex issue due to the fact that clinical manifestations of the AIDS do not represent a direct response of the organism to the harmful activity of HIV. In fact, infestation of T4 lymphocytes causes a deficiency of immune defense mechanisms, resulting in the undisturbed action of microorganisms that are, otherwise, harmless and not dangerous saprophytes.

It is also known that the immune defense mechanisms of the body exert significant dissuading effect on the development of neoplastic lesions[2]. Consequently, clinical manifestations of the AIDS depend on the microorganisms that develop pathogenic activity.

Incubation time of the AIDS appears to the broad. It is reported that four weeks after inoculation manifestations similar to infectious mononucleosis appear. Patients are presented with malaise, swelling of tonsils, ulceration of the nasopharyngeal mucosa and enlargement of neck lymph nodes. These clinical signs are present at the same time with the appearance of HIV antibodies in the serum of the patient. As we know, serum antibodies to HIV are detected at a time interval of 2-12 weeks after contamination.

The progression of HIV infection does not always lead directly to the AIDS. Often precursor states precede, such as lymph node syndrome (LAS) with persistent generalized lymphadenopathy (PGL)[3]. PGL or LAS is characterized by the appearance of lymphadenopathy in at least two focal points, beyond the inguinal region, lasting longer than three months. In these patients, the T4/T8 lymphocyte ratio is disturbed, and the patients are serum positive.

AIDS is characterized by the appearance of severe opportunistic infections, such as respiratory, central nervous system, or Kaposi's sarcoma. Clinical manifestations should be accompanied by detection of antibodies, HIV isolation in serum, detection of HIV antigens, isolation of the HIV from blood, and lymphocyte changes such as reduced numbers of T4 and T4/T8 cells.

The oral mucosa of these patients is a field for development of a variety of clinical manifestations with significant diagnostic, pathological and prognostic significance. Very often, oral manifestations are the first signs of the disease[4]. Oral manifestations are classified as microbial, viral, fungal, neoplastic or unwarranted. Depending on the frequency with which these manifestation occur in AIDS, they are classified as: Kaposi's sarcoma, Candidiasis, Non-Hodgkin lymphoma, and hairy leukoplakia.

Kaposi's sarcoma associated with AIDS appears in oral mucosa, face and upper torso. When the first cases of Kaposi's sarcoma were recognized as clinical symptoms of AIDS, it was easy for the clinicians to make the diagnosis due to the presence of sarcoma. With the advancement in the therapy of the disease, the Kaposi's lesions were not present in the majority of cases because the new and more successful treatments lowers the risk of the Kaposi's sarcoma.

Serum positive patients are vulnerable to Kaposi's sarcoma because their immune system is very weak. Also, HIV infected individuals with a CD4 count of less than 200 are at risk of developing Kaposi's sarcoma and are more likely to develop more altered forms of the diseases. Nonetheless, over the last 20 years we have seen that 40% of patients with Kaposi's sarcoma had a CD4 count of over 200. However, in patients with a CD4 count below 200, the risk is far greater and they are diagnosed with more heavy forms of the disease[8]. Kaposi's sarcoma occurs in seropositive patients receiving high CD4 antiretroviral therapy and detectable viral load. Cases like this have most often occurred in older men who have had HIV for many years and have spent some time in the part with a companion who has a very low CD4 count[9].

Kaposi's sarcoma lesions are lumps or nodules that might be red, purple, black or brown. They are mainly found in the oral cavity, skin, digestive and respiratory tract[10]. Intraoral lesions are primarily localized at the hard palate and secondly at the gingiva. The lesions in the mouth can easily be injured during mastication and this may cause bleeding or secondary infections.

Kaposi's sarcoma is not considered to be a real sarcoma. It originates from the mesenchymal tissue. Belonging to the neoplasms of the lymphatic epithelium, it carries vascular channels filled with blood cells. High vascular cells and intracellular beads are the hallmark of this tumor. Around the tumor there is a large swelling and pain due to inflammation. A definitive diagnosis can be made only by biopsy and microscopic examination that will show the presence of spindle cells. The detection of protein LANA of KSHV in the tumor cells confirms diagnosis[11].

Additional examination e.g. endoscopy or bronchoscopy are needed to confirm the presence of Kaposi's sarcoma in the digestive system or lungs[12]. An X-ray or radiographic examination (CT-Scan) might also be needed[13].

Kaposi's sarcoma is not easily cured, but is effectively mitigated for a long time and that is one of the treatment goals. When associated with immune deficiency, treating the causes of the immune system dysfunction may slow down or stop the development of the Kaposi's sarcoma. Techniques such as radiation and cryosurgery can be utilized to treat patients with local lesions.

In AIDS patients, Kaposi's sarcoma is considered to be an opportunistic infection, due to attenuation of the immune system. When Kaposi's sarcoma skin lesions appear, it is very likely that this is a sign of further weakening of the immune system.

Antiretroviral therapy is aimed at strengthening the immune system as well as slowing the progress of Kaposi sarcoma, thus extending the life of patients. Other classes of drugs targeting HIV are also effective against Kaposi's sarcoma. For many people with Kaposi sarcoma, this is the only treatment they need. This is seen in patients with CD4 above 150 and in limiting Kaposi's sarcoma lesions in the lymph nodes and skin. Over a period of several months the lesions should gradually shrink, fade and disappear.

The main goal in HIV therapy is the enforcement of the body defenses and that is lowering the risk of Kaposi's sarcoma appearance. Different drugs can have similar beneficiary result, and the therapy can be very effective for the overall therapy of the disease. Sarcoma lesions are controlled easier when the CD4 count is higher than 150, and when the lesions are in the skin and/or in the lymphatic system. After a period of several months, lesions usually shrink and gradually disappear[14].

Recommendation about treatment will be made by multidisciplinary team. This team usually consists of an oncologist, a dermatologist a pathologist and a dental surgeon that specializes in the treatment of oral cavity cancer.

Another widely used local treatment, injectable Vinblastine, is injected directly into the lesion. Radiotherapy (using high energy X-rays to destroy cancer cells) in an alternative technique. These techniques do not affect the development of lesions in untreated areas or use them for treatment in large areas. Nowadays new generation chemotherapeutic drugs are used. These include treatment with immunotherapy and targeted therapy drugs.

Conclusions

The doctor must be suspicious when lesions are observed, particularly in the oral cavity. A biopsy must be taken in order to obtain the correct diagnosis and examine the possibility of the presence of Kaposi's sarcoma or AIDS.

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