CASE REPORT

BACILLARY ANGIOMATOSIS IN AN ETHIOPIAN CHILD: A CASE REPORT

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ABSTRACT

We report a 9-year-old male HIV infected child who presented with multiple ulcerated lesions over the face and limbs of 6-month duration. With clinical suspicion of a malignant tumor, the child was admitted to Jimma University Specialized Hospital and a histopathologic evaluation suggested a diagnosis of Bacillary Angiomatosis .Consequently, he was treated with erythromycin and the lesions disappeared. This is a rare disease which is commonly associated with AIDS, usually occurring in adults. To the best of our knowledge, this is the first documented pediatric case in Ethiopia. The pathology, diagnostic and clinical features of this rare disease is discussed with a brief review of literature.

Key words: *Bacillary Angiomatosis, HIV, Bartonella*
Abbreviations*:- BA- Bacillary Angiomatosis HIV-Human Immunodeficiency Virus AIDS-Acquired Immunodeficiency Syndrome*

INTRODUCTION

Bacillary Angiomatosis (BA) is a recently recognized bacterial infectious vascular proliferative disease mainly seen in adult patients with AIDS. It was first described by Stoler et.al in 1983 in HIV positive male with multiple subcutaneous nodules (1). The disease usually manifests as cutaneous tumors but may have remarkable systemic manifestations. Involvement of the bone, spleen, liver, GIT, CNS, respiratory tracts, bone marrow, as well as bacteremia have been reported mainly with HIV-1 positive patients (2). The disease spectrum has also been expanded with reports in adults with other causes of immunosuppression and very rarely in immunocompetent individuals (3, 4). However for no clear reasons, BA is extremely rare in children. We describe this rare disease in a nine-year old male, HIV positive patient, and discuss the pathologic, clinical, and diagnostic features with a brief review of literature.

CASE REPORT

The patient is a nine-year old male from North-West Ethiopia who was a known stage IV AIDS patient for one year with a CD4+ count of 110/mm^3 at diagnosis. He was previously (2003) treated for disseminated tuberculous infection and repeated pneumonia attacks. In April 2008 (two months before the current presentation) the CD4+ count dropped to 80/mm^3, so HAART consisting of Zidovudine, Lamivudine and Efaviranz was started. But he was not on any other prophylactic therapy.

In March 2008, a swelling appeared over the right shin region which progressively increased in size and was associated with occasional pain. In two months, the lesions increased in number and were present over the left and right forehead, the chin, the right nostril, the right parietal region, the right flank region, and over the right distal radius. Eventually the lesion at the chin began to ulcerate and bleed. The child had intermittent low-grade fever, poor appetite and weight loss. He was the only child to his mother but has a 15 year old half elder brother through his father who was apparently healthy .The father was alive, reactive for HIV, apparently healthy, and on follow up at ART clinic. His mother died 4 years back after a diagnosis of tuberculosis. The family had a little monthly income earned by the father .There was a cat in the household with which the child had contact for a few years.

On physical examination, the child was emaciated with anthropometric measurements showing stunting and underweight. Vital signs were within the normal range; the pertinent findings were on the face where

there was a 4cm tumor over the chin with signs of necrosis, ulceration and oozing pus mixed with blood (Fig. 1) .Other lesions were found on the right parietal region (5x4 cm), right flank region (5x3 cm), right nose (2x2 cm), left forehead (4x3 cm)(Fig 2), right forearm (5 x 3 cm) (Fig.3) and right shin (5 x 3 cm). The lesions were mildly tender at presentation and slightly compressible on palpation with some mobility.

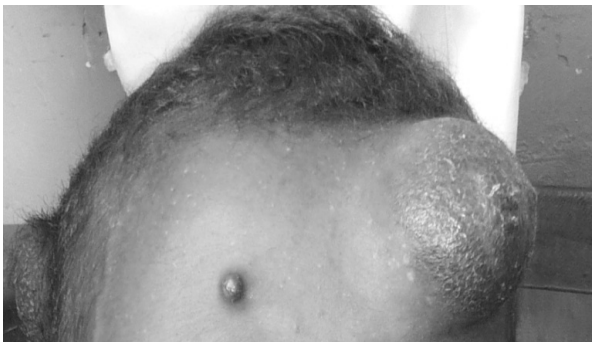

Fig 1: Multiple ulcerated and pus discharging lesions over the fore head and scalp at presentation

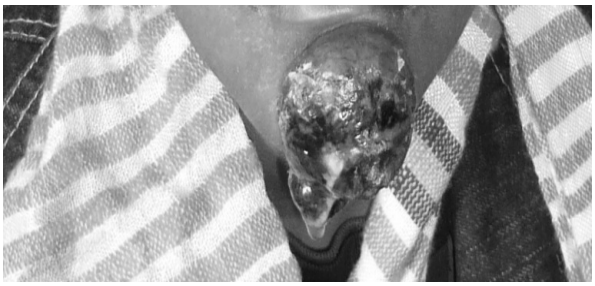

Fig 2: Ulcerated and pus discharging lesions over the chin

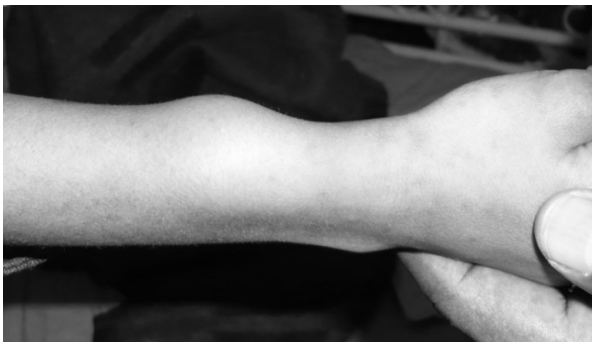

Fig 3: Right distal radial swelling at presentation

Chest X-ray showed no abnormal findings. The X-ray of the right tibia showed signs of periostal elevation and lytic lesions over the upper third. The hematocrit was 20 %, WBC count 5,600 with 56% neutrophils and 44% lymphocytes; platelet count was 125,000/mm3. The CD4+ count at presentation was 86/mm^3. Ultrasound evaluation of the abdomen was normal. Excisional biopsy was taken from the chin lesion and showed: chronic inflammation of the skin and subcutaneous tissue with slit-like vascular proliferation and a diagnosis of Bacillary angiomatosis was suggested (Fig 4).

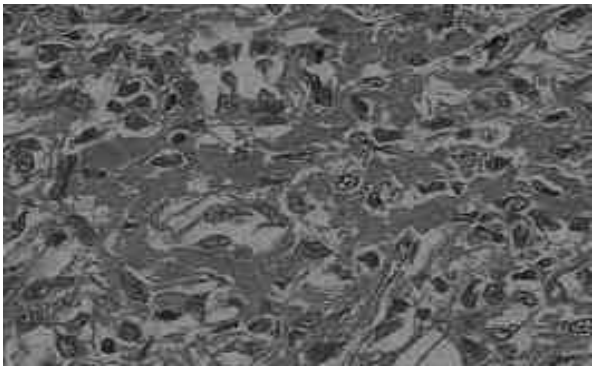

Fig 4: High power microscopic view showing slit like spaces and microvascular proliferation

The child was started on Erythromycin (250mg Po QID); after 2weeks of the treatment the lesions stopped to grow and no new lesion was seen. After a month of treatment, lesions started to decrease in size (Fig 5 and 6). The drug was continued for a total of 4 months after which the lesions disappeared totally and the child became well. After treatment, the CD4+ count increased to 285/mm^3, and X-ray of the right lower limb showed that the previous lesions completely disappeared.

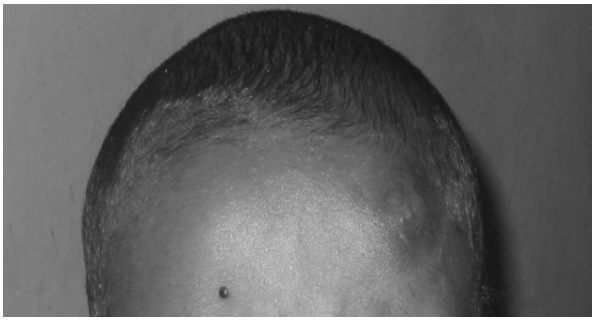

Fig 5: Disappearing lesions over the fore head one month after treatment

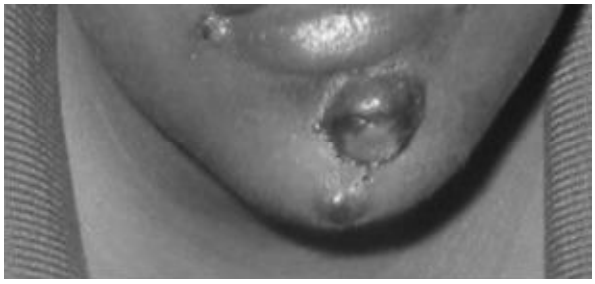

Fig 6: Disappearing lesions over the chin head one month after treatment

DISCUSSION

Bacillary Angiomatosis (BA) is a disease characterized by unique vascular lesions caused by infection with small, gram-negative organisms of the genus *Bartonella*. Almost all patients with this disease are HIV positive, but the disease range has subsequently expanded by reports of cases in other non-AIDS related immunosuppressive conditions and very rarely in immunocompetent individuals (5, 6).

The disease has different clinical spectrums which may be associated with liver, spleen, bone, and CNS diseases. BA occurs most frequently in the later stages of HIV infection; in a study conducted on 42 patients in the US, the median CD4+ cell count was 21 per cubic millimeter (range: 1-228) at the time of diagnosis of BA. In the same study, patients with BA infection presented with fever (9%), lymphadenopathy (21%), and/or abdominal symptoms (24%) (7).Our patient had no fever or lymphadenopathy at presentation; ultrasound examination didn't reveal any organomegally, either.

Two related bacteria of the genus *Bartonella, Bartonella heselae,* and *Bartonella quintana* are thought to be the causes of BA (8). *Bartonella heselae* infections are more frequently associated with the presence of lymphadenopathies. Subcutaneous and lytic bone lesions are strongly associated with *Bartonella heselae,* whereas peliosis hepatis is associated with *Bartonella quintana*. Epidemiological evidences suggest that *B. henselae* BA cases are more frequently associated with cat exposure, whereas those resulting from *B. quintana* are common among the homeless and dwellers in poor living conditions (9).

Cutaneous lesion of BA is usually an angiomatous papule or nodule resembling a pyogenic granuloma or a subcutaneous nodule with or without ulceration. Cutaneous nodules may be up to l0cm in diameter. BA may also present with systemic involvement, such as a destructive bone mass, lymphadenopathy, or colonic nodules without skin involvement. Symptoms may include weight loss and cough with or without haemoptysis or bloody diarrhoea (3, 10). In our patient, the presence of lytic lesion on the radius with periosteal elevation indicate the possibility of osteomyelitis like lesion by the same organism.

The diagnosis of cutaneous BA depends on histopathologic study. Skin specimens can be obtained by shave excision or punch biopsy. Tissue specimens reveal a characteristic vascular proliferation on routine hematoxylin & eosin staining. There is a typically lobular proliferation of small, capillary-sized blood vessels with protuberant, cuboidal, or polygonal endothelial cells containing abundant cytoplasm, with or without cytologic atypia. In addition numerous bacilli are demonstrable by modified silver staining (eg, Warthin-Starry, Steiner, Dieterle) or electron microscopy. We couldn't perform the above stainings on our specimen for technical limitations in the lab. Histologically, BA can be confused with other vascular tumors like Kaposi Sarcoma ,angiosaroma and pyogenic granuloma(11,12).

Although BA predominantly occurs in adult AIDS patients, there are reports in children and immunocompetent individuals. Cockerell CJ et.al. described the disease for the first time in a 37-year old healthy man without any apparent immunocompromizing illness, and concluded that BA can occur in immunocompetent individuals(6,13). Smith et.al. described the disease in an immunocompetent 6-year old girl after she was found to have red papular lesion over the neck region; however, for unknown reasons BA is less common in children than adults (15).

Antibiotics are the main modes of treatment of BA. Erythromycin and Doxycycline are the two important first line drugs which were found to be effective in treating many patients with BA (5, 9, and 15). If these antibiotics are not tolerated, tetracycline, clarithromycin, or azithromycin can be used. The initial BA patient described by Stoler in 1983 was treated empirically with erythromycin and had complete resolution of lesions (1). In case of cutaneous BA before initiation of treatment, a complete evaluation should be done for parenchymal or osseous involvement. Cutaneous BA usually treated with oral antibiotics and response is often dramatic. Improvement can occur in a week, and in one month a complete resolution can be seen, but many recommend continuing treatment for 8-12 weeks (16). In case of involvement of the liver or bone, longer treatment up to 4 months is recommended (17).Because of systemic involvement in our patient, we treated him with Erythromycin 250mg Po QID for a total of four months with excellent results.

In conclusion, BA is a rare disease which commonly occurs in adult AIDS patients. Our case and other case reports call attention to the occurrence of the disease in the pediatric age group .We believe physicians should consider BA in their list of differential diagnosis of cutaneous and systemic tumours in AIDS patients in any age group. Accordingly,

prompt histopathologic evaluation must be made for correct diagnosis since the disease can be effectively cured with commonly available antibiotics.

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